Contents

PREFACE

The first, second third and forth edition of Manual of Clinical Pediatrics proved to be very successful as a practical manual, both here and overseas; providing the opportunity for this fifth edition.

Many chapters in the fifth edition of this manual have been extensively updated and revised. Immunization chapter and intensive care chapter is updated in this edition. However, the format of the manual has remained essentially the same.

The management principles and protocols in the update of this manual are not based on practices at only one institution. Contributors are from different institutions in the Kingdom of Saudi Arabia. I feel this has enabled us to produce a manual that is not institution-specific but reflects a cross section of contemporary approaches to pediatric management.

My sincerest thanks to the contributors of the first, second, third, fourth and fifth edition, and my family for their continued support, which has been indispensable to the completion of this edition.

I would like to extend my **special thanks to Dr. Sameeh Ghazal** who had worked very hard, in spite of his tight schedule on reviewing all the manuscripts, in addition to his contribution to the manual by two chapters, several drawing, and figures.

I welcome any suggestions and comments regarding the manual; letters should be addressed to:

Dr. Mansour Al-Howasi
P.O. Box 62300
Riyadh, 11585
K.S.A.

CONTRIBUTORS
(Alphabetic order)

- **PROFESSOR ABDULLAH AL HERBISH**
MBBS, FRCPC, FAAP
Consultant, Professor of Pediatrics and Endocrinology
College of Medicine & King Saud University, Riyadh
(Endocrinology)

- **Dr. ABEER AL MOLLA**
MBBS, Fellowship in Radiology
Consultant in Radiology
King Fahad Specialist Hospital
Dammam, KSA
(Radiological Imaging)

- **Dr. ALIA AL IBRAHIM**
MBBS, DCH, CABP, Fellowship in Paediatric Nephrology
Consultant in Paediatrics & Paediatric Nephrologist
KKUH Hospital
Riyadh, KSA
(Renal System)

- **PROFESSOR ASAAD ABDULLAH ASSIRI**
MBBch, MRCP (UK), FRCP
Consultant, Professor of Paediatrics and Gastroenterology
King Saud University, Riyadh, KSA
(Gastrointestinal System)

- **Dr. DHIMAN CHOWDHURY**
MBBS, DCH, MRCP (UK), FRCP (Edin.)
Consultant and Assistant Professor of Paediatrics
IWK Grave Health Center, Department of pediatrics, Dalhousie University. Canada
(Gastrointestinal System)

- **Dr. IBRAHIM S. ALHIFZI**
MBBS, DCH, CABP, FRCPCH, FRCP
Consultant Neonatologist
Clinical Professor of Paediatrics
Armed Forces Hospital, Southern Region
(Examination of Neonate)

- **Dr. MAHER AHMED KHALIFA**
MBBch, DCH, MRCP (UK)
Consultant Paediatrician & Paediatric Neurologist
Insurance Hospital (Riyadh), Riyadh, KSA
(Neurology & Development)

- **Dr. MANSOUR M. ALQURASHI**
MBBS, DCH, MRCP (UK), ABP, Fellowship in Cardiology
Consultant Paediatrician & Paediatric Cardiologist
AlYammamah hospital,
Clinical Assistant Professor of Paediatrics
King Saud University
Riyadh, KSA
(Cardiovascular System)

- **Dr. MANSOUR NASER ALHOWASI**

MBBch, DCH, MRCP (UK), FRCP (Edin.)
Consultant Paediatrician
King Fahad Medical City, Children's Hospital
Clinical Assistant Professor of Paediatrics
King Saud University
Deputy Minister for Executive Affairs, MOH, Riyadh, KSA
(History and Clinical Examination, Neonatology, Immunization, Nutrition and infant feeding, Cardiovascular system, Gastrointestinal system, Urinary system, Neurology and development, Infectious Diseases)

- **Dr. MOHAMMED F. FAROUQ**

MBB.ch, FAAP
Consultant in Paediatrics and Infectious Disease
Assistant Professor in Paediatrics and Haed of paeditrics department
King Abdulaziz University, Jeddah, K.S.A.
(Immunization)

- **PROFESSOR MOHAMMED MOHAMMED JAN**

MBBch, FRCPC
Consultant Paediatric neurology
Professor of Paediatrics & Neurology
King Abdulaziz University Hospital &
King Faisal Specialist Hospital and Research Center, Jeddah, K.S.A.
(Neurology)

- **Dr. SAMEEH S. GHAZAL**

MBBch, DCH, MRCP (UK), ABP, MRCPCH (UK), CIC (USA)
Consultant Paediatrician, Infectious Diseases and Infection Control
King Fahad Medical City, Children's Hospital
Clinical Assistant Professor of Paediatrics
King Saud University, Riyadh, KSA
(Infectious Diseases & Respiratory System and Bronchial asthma)

- **Dr. SMRITI CHOWDHURY**

MBBS, DCH, MRCP (UK)
Consultant Paediatrician
Department of neonatology, Dalhousie University. Canada
(Gastrointestinal System)

- **Dr. WAFAA AL SUWAIRI**

MBBS, DCH, MRCP (UK), ABPP.
Consultant Paediatric Rheumatologist
King Abdulazaz Medical City, National Guard, Riyadh, K.S.A.
(Musculoskeletal System)

- **Dr. WALEED HAMED ALBUALI**

Consultant pediatric intensivist, Assistant professor
Pediatric ICU director, Department of Pediatrics
King Fahd Hospital of the University
Al-khobar, Saudi Arabia
(Emergency & Intensive Care)

- **Dr. YOUSEF K. GHAZAL**

MBBch, MRCP (UK), CABP, SBP, MRCPCH (UK)
Consultant Paediatrician, Intensivest
King Fahad Medical City, Children's Hospital, Riyadh, KSA
(Nutrition & Infectious Diseases)

HISTORY & CLINICAL EXAMINATION

HISTORY

- Name:
- Age (Date of birth):
- Sex:
- Nationality:
- Address:
- Source of history:
- (mother, father & others)

PRESENTING COMPLAINTS:

- Use their own words. (The parents may actually tell you the diagnosis).
- Symptoms with duration

HISTORY OF PRESENTING COMPLAINTS:

- Obtain a complete chronological sequence of events. Deeper inquiry about important symptoms must be made regarding:
  - Onset
  - Course
  - Duration
  - Site
  - Frequency
  - Severity
  - Relieving factors
  - Exacerbating factors
  - Diurnal or seasonal variation
  - Relation to food
  - Relation to exercise e.g. cough
  - School missing related to the complaint

  *Any associated symptoms

SYSTEM INQUIRY:

System inquiry is important when there is complain not specific for one system. e.g. fever or when there is multisystem disease. There are few questions, which provide useful "screening"

General: e.g. - Feeding and appetite (very important)
- Irritability
- Weight loss

Cardiovascular: e.g. - Breathlessness
- Sweaty on feeding
- Cyanosis

Respiratory: e.g. - Breathlessness
- Runny nose

- Cough
- Noisy breathing (wheeze or stridor)
- Sore throat or earache
- Hemoptysis

Gastrointestinal: e.g.- Vomiting
- Abdominal pain
- Constipation or diarrhea (frequency and appearance of stool)
- Jaundice

Genitourinary: e.g. - Frequency
- Dysuria
- Nocturia or enuresis
- Hematuria
- Incontinence
- Age of menarche

Neurological: e.g. - Irritability
- Drowsiness
- Fits or abnormal movements
- Headaches
- Numbness or unpleasant sensation
- Weakness

Hematological & Oncological: e.g.
- Pallor
- Jaundice
- Bone pain
- Bruises
- Bleeding from the nose

Infections: e.g. - Skin rash
- Contact with infectious patients
- Recent travel

Musculoskeletal & skin: e.g.
- Joint swelling
- Joint pain
- Skin rash

N.B. If any symptom during the system review is positive, deeper inquiry must be made:
e.g. Cough - Nocturnal or related to exercise points towards b. asthma. Purulent sputum points towards suppurative lung disease.

PAST MEDICAL HISTORY:

These include:
- Previous disease
- Previous medications taken by the patient:

- Frequency - Dose
- Previous hospitalization
- Previous surgery
- Previous transfusion
- Any known drug or food allergies?

PREGNANCY AND NEONATAL HISTORY:

- Follow up during pregnancy
- Mother's illness during pregnancy (nature of the illness- which trimester) e.g. - flue like illness or skin rash during early pregnancy may point towards congenital infections.
- Mother's medication: e.g. - Valproate taken by the mother during pregnancy increase risk of fetal neural tube defects.
- Anabolic steroids taken by the mother during pregnancy may cause virilization of female fetus.
- Phenytoin taken by the mother during pregnancy may increase risk of fetal congenital anomaly especially cardiac anomaly.
- Exposure of the mother to radiation during pregnancy.
- Fetal movement: Weak fetal movement may point towards intrauterine hypotonia. e.g. - Dystrophia myotonica, spinal muscular atrophy.
- Polyhydramnios: - May point towards fetal GIT obstruction. e.g. Esophageal atresia or intrauterine hypotonia.
- Oligohydramnios: - May point towards fetal urinary system abnormality. e.g. bilateral renal agenesis.
- Length of gestation.
- Mode of delivery.
- Birth weight, height and head circumference.
- Apgar score.
- Any neonatal disease or admission and why?

NUTRITIONAL HISTORY:

- Breast-fed or bottle-fed and for how long?
- If bottle-fed:
    - Which formula did he receive?
    - How was it prepared?
    - What volume did he take at each feed?
    - And how long did he take it?
    - Frequency of feeds
    - Total daily intake
- Time of weaning - timing of introduction of solids and cereals.

N.B. Normally, breast-fed baby might pass up to 6 motions daily.

IMMUNIZATION HISTORY:

The recommended vaccination in Saudi Arabia described later on:

- Check immunization card.
- If there is failure in taking the vaccine ask for the reasons in details.

DEVELOPMENTAL HISTORY:

- This includes:
    - Gross motor
    - Fine motor
    - Visual
    - Speech and hearing
    - Social and play
    - Schooling (level and performance)

N.B. - If the mother has other children, compare his or her development with his or her other siblings.
- Some important development milestones have been described later on.

FAMILY AND SOCIAL HISTORY:

- Ages of parents
- Consanguinity
- Number of siblings and age range (any sibling from previous or another marriage)
- Family history of similar condition
- Which region the parents originally came from (e.g. sickle cell anemia common in south-west and eastern region of Saudi Arabia)
- Neonatal deaths (e.g. metabolic disease)
- Previous abortions
- Housing - type of accommodation (rented or owned, house or flat, number of bedrooms, washing and toilet facilities, air conditions and heaters)
- Parents' occupation and income of the family
- Parents' education
- Parents' smoking habit (especially in bronchial asthma cases)
- Contact with animals
- Recent travels (e.g. malaria in southwest of Saudi Arabia)

N.B.

- Pay more attention to detailed family history, if hereditary, allergy or infectious disease is involved e.g. sickle cell anemia, bronchial asthma, and tuberculosis.
- Transportation: Make sure there is available transportation for the child and his or her attendance to be able to attend any follow-up.
- Try to make the appointment of any follow up suitable for the condition of the father's work.
- If the patient came from poor family, contact the social worker to arrange for the family financial support and airplane tickets between the regions if the family came from far area...etc.
- Golden role: At the end of your history, ask the historian if she or he likes to inform you anything else or if she or he expect you to ask her or him any other question about her or his child that was not asked yet.

GENERAL ADVICE ON EXAMINING CHILDREN

1. Introduce yourself to the child and or his attendant.
2. Ask the child's name.
3. Wash your hands especially if the child is an infant.
4. Warm your hands and remove watch or ring, which might scratch the child.
5. Inform the child that you are going to examine him e.g. I am going to percuss your chest
6. Avoid standing over a small child by getting down to his level.
7. Distract the child with a toy or any other thing if this will help you to continue your examination.
8. Talk to the child as you examine and smile to him or her.
9. Remember to thank him at the end of the examination.
10. During examination some of the examiners like to hear running commentary while you examine the child.

"NEVER & DO NOT"

- Never handle a child roughly.
- Never refer to a child as dysmorphic without first seeing the parents (except the common known syndromes e.g. trisomy 21).
- Do not get the sex of the child wrong.
- Do not use potentially worrying terms in front of parents without explaining them e.g. tumor or mental retardation.
- Do not use abbreviations in your clinical notes except if it is internationally known and accepted.
- Do not discuss the case with your colleague in front of parents using foreign language without explaining to the parents what you are doing and reassuring them.

CLINICAL EXAMINATION

Vital signs:

Growth Parameter:

- *Weight*
- Height
- Head circumference

(Plot them on standard centile chart)

Inspection:

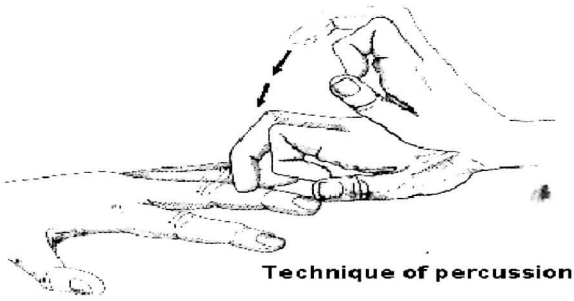
A great deal of information can often be elicited without even touching the child. In addition some children will cry as soon as you touch them. You should expose the relevant area, (the whole chest, the whole abdomen or the legs). If the parents are present ask them to undress the child, otherwise you will miss an operation scar, hydrocele, muscle wasting or some other important signs. Do comment on the general condition of the patient (well or ill), and on intravenous drips, nasogastric tubes, urinary catheter, or obvious dysmorphic features if present...etc.

Palpation:

If there is any possibility of the part you are palpating being painful, you should ask the patient if it hurts and be particularly gentle.
e.g. abdomen, joints and lumps.

Percussion:

- Do not forget that percussion is an important method to detect organomegaly especially in young children.



Technique of percussion.

Auscultation:

- Do not forget to auscultate the abdomen for intestinal sound and bruits
- Do not forget to auscultate the anterior fontanelle for bruits when you examine a newborn with heart failure. (A-V malformation)
- N.B. - Try to start from the periphery then go central
- Sequence of examination might be difficult to apply in young children

GOLDEN RULES

- **If there is one congenital anomaly, look for other congenital anomalies**
  e.g. examine the heart and urinary system in any child with dysmorphic features
- **If there is one endocrine disease, look for other endocrine diseases**
  e.g. look for signs of hypothyroidism in any diabetic patients
- **If there is one nutritional disease, look for other nutritional disease**
  e.g. look for iron deficiency anemia in any patient with rickets
- **If there is one "atopic" diseases look for other atopic diseases**
  e.g. look for atopic eczema in any patient with bronchial asthma
- **If there is one autoimmune disease, look for other autoimmune diseases.**
  e.g. look for signs of Addison's disease in any patient who has alopecia areata or vitiligo

65 of infant motality of in Neoborn peroid.

NEONATOLOGY

The success of the health care system in countries is commonly judged by infant mortality rate (death occurring from birth to 12 months / 1000 live births). The neonatal period (1st four weeks of life) is a highly vulnerable time for the infant and neonatal mortality where it accounts for about 65% of infant mortality; hence proper care of the neonate contributes significantly to reduction in infant mortality.

PT, DM, oligh polyh, PROM infect, HPN Fetal dist.

I. Delivery Room Management

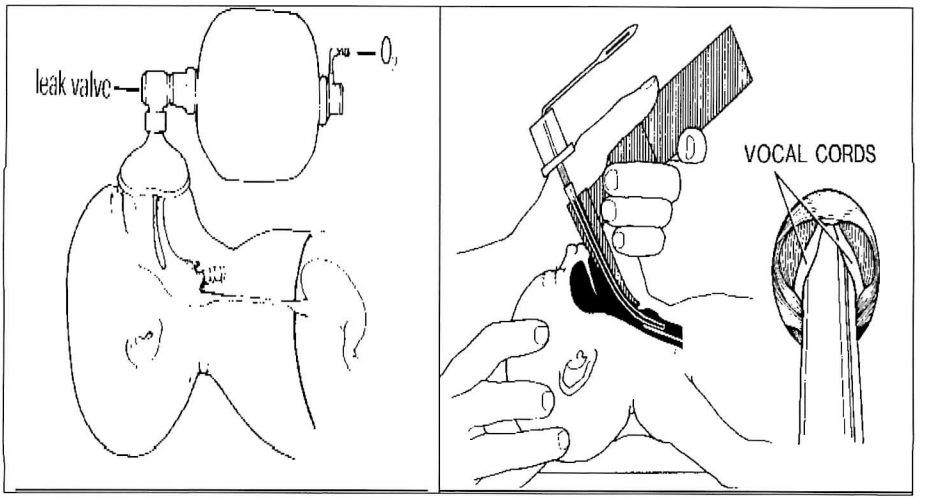
1. Anticipation

Proper information obtained from the obstetrician or the midwife about maternal condition should identify neonates at risk of developing problems in the delivery room. This includes mothers with certain diseases or problems such as premature labor, IUGR, prolonged rupture of membranes (18 hours before delivery), oligohydramnios, polyhydramnios, diabetes, pregnancy induced hypertension, infections, fetal distress, etc.

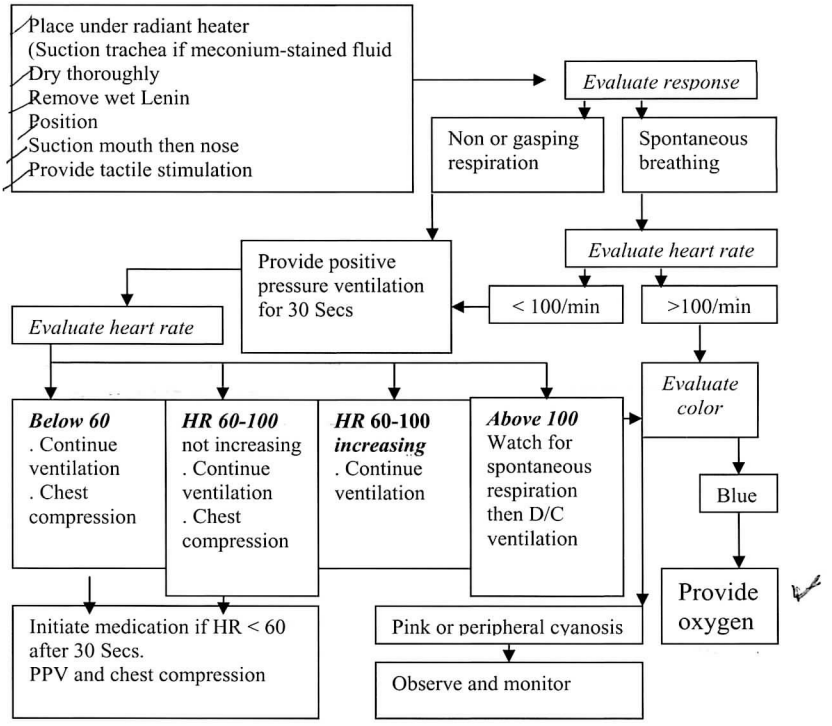
2. Preparation

Check the resuscitation equipment and the medications needed for resuscitation.

1. Radiant warmer switched on, warm towels
2. Ambu bag connected to air-oxygen blinder
3. Laryngoscope and blades (Size: 00, 0, 2)
4. Endotracheal tubes (size 2.5, 3, 3.5)
5. Stethoscope
6. Stop clock
7. Catheters, cannulas, syringes and needles
8. Medications



3. Resuscitation:



- Resuscitation with room air is safe and effective
- Intrapartum suctioning of oro and nasopharynx is not recommended for infant born with meconium stained liquor and suction of trachea is not recommended if baby is vigorous.
  During this process Apgar score should be estimated as follows:

SCORE	0	1	2
A = Appearance	Blue / Pale	Blue extremities	Pink
P = Pulse (HR)/min.	0	< 100	> 100
G = Grimace (Reflex Irritability)	None	Grimace	Cry
A = Activity (Muscle Tone)	Flat	Some limb flexion	Active movements
R = Respiration	Absent	Slow, irregular	Strong cry

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Chest compression techniques (cardiac massage)

A) Two Thumbs Method (Preferred)
Place both thumbs on the middle third of the sternum just below an imaginary line drawn between the nipples, with the fingers encircling the chest and supporting the back. The xiphoid or lower portion of sternum should not be compressed to avoid abdominal trauma.

B) Two Finger Method
The index and middle finger are placed over the middle third of the sternum.

Epineph.
Nalaxone 0.1mg/kg
ET dose → 0.1 mg/kg
IV → 0.01mg/kg

Medications
The medications that are currently recommended during resuscitation of the newborn infant are:
Adrenaline (Epinephrine): 0.01 - 0.03 mg / kg of 1:10000 solution (using high dose Epinephrine may lead to hypertension, decreased myocardial functionand poorneurological outcome. ET dose is 0.1 mg/kg/dose.
Naloxone: Used to reverse respiratory depression caused by narcotic administration to the mother. Dose: 0.1 mg / kg Route: IV only.
Volume Expander: 10 mls / kg as Normal Saline, or O-Negative blood if blood loss is suspected.
The routine use of Sodium Bicarbonate is discouraged, except in prolonged arrest. Atropine and calcium is not recommended in resuscitation of the newborn.
A brief examination should be done in delivery room to look for major congenital malformations. The baby will then be sent to Nursery or Neonatal ICU according to his/her condition.

II. CARE OF THE NORMAL NEWBORN*

Normal newborns should be given to mothers as soon as they are stable enough to be breast-fed and should always be nursed with their mothers. A detailed examination must be performed within 24 hours of birth. It should be done in the presence of the mother or both parents to answer their questions and to give advice about feeding and care of their baby. Vaccinations should be given, routinely BCG and hepatitis B vaccine, but hepatitis B immunoglobulin is added to babies of mothers with hepatitis B positive screening.

PHYSICAL EXAMINATION OF THE NEWBORN INFANT

This requires patience, gentleness, and flexibility, so auscultation of the heart and palpation of the abdomen can be done while the infant is quiet and relaxed. Adequate light, warm hands and environment are prerequisite. Proper hand washing before examination is essential.

- Vital signs should be recorded: Pulse (normal 120 - 160 / minute), Respiratory Rate (30 - 60 / minute), and temperature.
- Weight, length, and head circumference should be plotted on Centile chart.

GENERAL APPEARANCE

- Alertness
- Movements
- Color (cyanosis of cold periphery is normal)
- Dysmorphism

SKIN

- Pallor (circulatory failure or anemia)
- Mottling
- Plethora (polycythemia)
- Jaundice
- Birth marks, hemangiomas, mongolian blue spots
- Rash (erythema toxicum, septic spots, herpes, transient pustular melanosis)
- Edema (generalized: hydrops) (localized: hands and feet in Turner's syndrome)

HEAD

- Size
  - Microcephaly: familial, congenital infection
  - Macrocephaly: hydrocephalus, familial, achondroplasia,hydranencephaly, etc.
- Shape
- Fontanelles & sutures
- Masses - cephalohematoma (collection of blood under the periosteum which does not cross sutures) - Caput succedaneum (edematous scalp of the presenting part)

FACE

- Dysmorphic features

EYES

- Microphthalmia as in congenital rubella syndrome
- Buphthalmos (corneal diameter > 1 cm)
- Slant of palpebral fissures (upward or downward)
- Conjunctival hemorrhage
- Coloboma of the lids or the iris (syndromes)
- Aniridia (association with Wilms tumor and other urogenital anomalies)
- Red reflex suggests absence of cataract and major intraocular pathologies
- Leukokoria (white pupillary reflex) seen with cataract, ROP, and retinoblastoma
- Hypertelorism (widely spread eyes), or hypotelorism

EARS

- Low set ears (classically seen in Down's Syndrome)
- Malformations and periauricular tags
- Ear drums appear normally dull gray

Large tongue
- Hypothyroid
- Beckwith-Wiedemann syn.

MOUTH
- Natal teeth (remove only if loose or interfere with feeding)
- Large tongue is seen in congenital hypothyroidism, Beckwith-Wiedemann syndrome and others but glossoptosis is seen with Pierre Robin syndrome
- Cleft lip and palate
- High arched palate
- Lingual thyroid

NECK
- Normally short
- Swellings: goiter, cystic hygroma, thyroglossal cyst, sternomastoid mass.
- Redundant skin at the back of the neck is seen with Down's Syndrome and is associated with webbing in Turner's Syndrome

RESPIRATORY
- Chest Shape: pectus, nipples (engorgement, space) movements
- Sounds: (grunting, stridor, crying), air entry, breath sounds, added sounds

CARDIOVASCULAR
- Pulse: rate, rhythm, volume, etc.
- Weak femoral pulse and higher BP in upper limbs than lower limbs (Coarctation of the Aorta)
- Dextrocardia
- Heart sounds
- Murmur

ABDOMEN
- Distended or scaphoid
- Organomegaly
- Hernias
- Bowel sounds

GENITALIA
- Sex
- Ambiguity
- Testis
- Labial fusion
- Hypospadias
- Check anal patency

NEUROLOGY
- Alertness
- Movement and posture
- Tone
- Reflexes
- Primitive reflexes: (Leave them to the end of examination)

Reflex	*Stimulus*	*Response*	*Appearance (Gestational age)*	*Disappearance (Corrected age)*
Moro	Lift the head slightly and then drop it gently on your palm	Rapid extension & abduction of arms with hand opening followed by slow return to mid line	28 – 32 weeks	3 – 5 months
Sucking	Nipple or teat	Strong & synchronized sucking	32 weeks	4 – 7 months
Rooting	Gently stroke the cheek with finger tip	Baby searches with his mouth	32 weeks	4 – 7 months
Palmar	Touch the baby's open palm with your finger	Baby grasps the finger	32 weeks	3 – 4 months
Plantar	Firmly press the ball of infant's sole with your thumb	Toes flex	32 weeks	8 months
Stepping	Hold the infant upright with the sole touching a flat surface	Alternating stepping movement	34 weeks	2 months
Placing	Touch the dorsum of a foot with the edge of the table	Baby climbs over the edge of the table	34 weeks	5 months

EXTREMITIES
- Size and shape (e.g. hemihypertrophy)
- Digits (count, syndactyly, etc.)
- Club feet

BACK
- Mongolian blue spots
- Haïr tuft
- Dimples
- Sinus
- Scoliosis

HIPS
- Asymmetry of groin creases
- Barlow's & Ortolani's Test - (to assess hip dislocation)
  Lie the infant supine on a flat hard surface and remove the nappy. Stand in the mid line at the foot end of the infant, flex infant's knees fully and hips to 90°. Hold the lower limbs with your thumbs on the medial condyles and tips of the middle finger

on the greater trochanters of each femur. Bring the knees together and attempt to push the hips posteriorly. If you feel a 'click', it means the head of the femur has dislocated (Barlow's sign). Keeping the grip unchanged, now abduct the infant's thighs with the thumbs and lift the femoral heads forward with the middle fingers. If you feel a definite 'clunk', it means the previously dislocated head of the femur has slipped back into the acetabulum (Ortolani's sign).

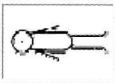
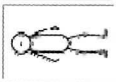
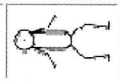
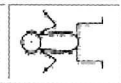
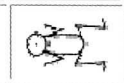
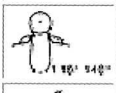
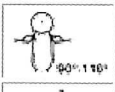
THE PREMATURE NEONATE

The New Ballard Score

New Ballard Score Sheet

Use this score sheet to assess the gestational maturity of your baby. At the end of the examination the total score determines the gestational maturity in weeks.

NEUROMUSCULAR MATURITY

SIGN	SCORE							SIGN SCORE
	-1	0	1	2	3	4	5	
Posture								
Square Window								
Arm Recoil								
Popliteal Angle								
Scarf Sign								
Heel To Ear								
TOTAL NEUROMUSCULAR SCORE								

PHYSICAL MATURITY

SIGN	SCORE							SIGN SCORE
	-1	0	1	2	3	4	5	
Skin	Sticky, friable, transparent	gelatinous, red, translucent	smooth pink, visible veins	superficial peeling &/or rash, few veins	cracking, pale areas, rare veins	parchment, deep cracking, no vessels	leathery, cracked, wrinkled	
Lanugo	none	sparse	abundant	thinning	bald areas	mostly bald		
Plantar Surface	heel-toe 40-50mm: -1 <40mm: -2	>50 mm no crease	faint red marks	anterior transverse crease only	creases ant. 2/3	creases over entire sole		
Breast	imperceptable	barely perceptable	flat areola no bud	stippled areola 1-2 mm bud	raised areola 3-4 mm bud	full areola 5-10 mm bud		
Eye / Ear	lids fused loosely: -1 tightly: -2	lids open pinna flat stays folded	sl. curved pinna; soft; slow recoil	well-curved pinna; soft but ready recoil	formed & firm instant recoil	thick cartilage ear stiff		
Genitals (Male)	scrotum flat, smooth	scrotum empty, faint rugae	testes in upper canal, rare rugae	testes descending, few rugae	testes down, good rugae	testes pendulous, deep rugae		
Genitals (Female)	clitoris prominent & labia flat	prominent clitoris & small labia minora	prominent clitoris & enlarging minora	majora & minora equally prominent	majora large, minora small	majora cover clitoris & minora		
							TOTAL PHYSICAL MATURITY SCORE	

MATURITY RATING

TOTAL SCORE (NEUROMUSCULAR + PHYSICAL)	WEEKS
-10	20
-5	22
0	24
5	26
10	28
15	30
20	32
25	34
30	36
35	38
40	40
45	42
50	44

References:

Ballard JL, Khoury JC, Wedig K, *et al*: New Ballard Score, expanded to include extremely premature infants. *J Pediatrics* 1991; 119:417-423

Complications

The more immature the neonate the severer and more frequent the complications

- Respiratory distress syndrome
- Apnea
- Intra Ventricular hemorrhage
- Jaundice
- Metabolic derangements
- Infections
- Necrotizing enterocolitis
- Retinopathy of prematurity
- Chronic lung disease
- Rickets
- Anemia

SOME NEONATAL PROBLEMS OF INTEREST

Birthmarks:

- **Mongolian spots -** These are blue or gray macules usually seen over the lumbosacral region or buttocks. Commonly seen in Asian races. Most of them regress spontaneously by 2 - 5 years.
- **Salmon patch -** These are red macules usually seen over the nape of the neck, eyelids and forehead (stork bite). These are present since birth and mostly fade by one year except those on the nape of the neck.
- **Strawberry nevus -** These are raised red lesions with well-defined borders. Most of them are not present at birth, but appear within first two months. These increase initially in size and then regress spontaneously by the age of two years.
- **Port wine nevus -** These are red or purple macular lesions, which are present since birth and don't show any regression. Involvement of the face may be associated with meningeal hemangiomatosis (Sturge-Weber syndrome).

Erythema toxicum:

These are erythematous maculopapular rashes most marked over the trunk and usually develop in the first few days of life. These lesions disappear spontaneously within a few days.

Milia:

These are yellowish-white specks seen mostly over the cheeks and nose, which consist of minute sebaceous cysts. These are usually seen in the first few days of life and disappear spontaneously within 3 - 4 weeks.

Miliaria:

These are crops of erythematous papules or papulo-vesicles mostly seen over the forehead, neck and napkin area in infants nursed in a warm and humid atmosphere. These lesions generally resolve on exposure to a cool environment.

Breast engorgement:

Breast swelling and redness in the newborns of either sex with slight milk discharge (witch's milk) is common and is due to transient stimulus of maternal estrogen. It resolves spontaneously within a few weeks. The breast should not be squeezed as it may introduce infection.

Caput succedaneum:

It is a diffuse edematous swelling of the scalp over the presenting part of the head during delivery. The swelling extends across the suture lines. It is present at birth and disappears spontaneously within the first few days of life.

Cephalhematoma:

This is a localized swelling of the scalp usually seen over the parietal area due to subperiosteal hemorrhage and is limited by the suture lines. It may be bilateral. The swelling is usually not visible until several hours after birth. It resolves spontaneously within two weeks to three months. It may be complicated by significant jaundice and anemia. It should not be interfered with unnecessarily as this may introduce infection.

Umbilical hernia:

It is a swelling at the base of the umbilicus due to protrusion of the omentum or small intestine through the fibrous ring. The swelling is covered by skin and is easily reducible. Strangulation is extremely rare. Most umbilical hernias disappear spontaneously by 1 - 2 years.

Omphalocele:

It is also a herniation of abdominal contents into the base of the umbilical cord. The sac is covered with peritoneum without the overlying skin. It is often associated with Beckwith-Wiedemann syndrome. Immediate surgical repair is needed for this condition.

Gastroschisis:

It consists of a complete defect through all the layers of the anterior abdominal wall usually to the right side of the umbilical cord through which the gut prolapse. In contrast to omphalocele it has no covering and the umbilical cord is normal. This is a surgical emergency and the patient has to be transferred immediately for surgery in cellophane wrapping as the infant loses fluid and heat readily from the exposed gut.

Inguinal hernia:

It usually presents as a swelling at the external inguinal ring, which may extend down into the scrotum or labia majora most marked when the infant is straining. In contrast to umbilical hernia it needs early surgical repair as the chances of strangulation is high in younger age.

Hydrocele:

The hydrocele, in contrast to the inguinal hernia, is more marked in the dependent part of the scrotum; the upper limit of it can be reached and transillumination is positive. Small hydrocele disappears by one year, but larger ones require surgical intervention.

Erb's palsy:

It results from injury to upper brachial plexus (C5 and C6) due to excessive traction on the neck at delivery. It usually follows difficult deliveries. The infant assumes the *'waiter's tip position'* with adduction and internal rotation of the arm, pronation of the forearm and flexion of the wrist joint. The

recovery is usually complete by 6 months, mostly within the first month, but some may extend up to 18 months.

Neonatal jaundice:

Jaundice is a common neonatal problem with an incidence of 60% in term and 80% in preterm infants. It is mostly due to physiological jaundice, which starts after 24 hours of life and fades away by one week.

N.B. Jaundice developing within first 24 hours of life is always pathological.

Evaluation of neonatal jaundice:

History:

Importance should be given to the following points in the history:

- Onset of jaundice
- Family history of neonatal jaundice (breast milk jaundice, hemolytic anemia, hereditary non-hemolytic anemia)
- Flu-like illness or skin rash during pregnancy (congenital infections)
- Type of feeds - breast or bottle
- Blood group of mother and baby

Examination:

Look particularly for the following signs (which might point to the etiology) -

- General - dysmorphic features (Alagille syndrome, Zellweger syndrome), obese and plethoric (infants of diabetic mother)
- Head - microcephaly (congenital infections), cephalohematoma, large fontanelle (cretinism)
- Eyes - cataract (galactosemia), chorioretinitis (congenital infections)
- Mouth - large protruding tongue (cretinism)
- Skin - bruises, purpura, petechiae (congenital infections)
- Abdomen - hepatosplenomegaly (congenital infections, hemolytic anemia)
- Umbilicus - infection, hernia (cretinism)

Causes of neonatal jaundice:

Indirect hyperbilirubinemia: (Direct serum bilirubin < 15%)	*Direct hyperbilirubinemia:* (Direct serum bilirubin = 15%)
- Physiologic jaundice	- Congenital infections
- Breast milk jaundice *	- Sepsis
- Blood group incompatibilities	- Neonatal hepatitis*
- Urinary tract infection *	- Biliary atresia *
- Internal hemorrhage	- Choledochal cyst *
- Infants of diabetic mother	- Inspissated bile syndrome *
- G-6-PD deficiency	- Galactosemia *
- Hereditary spherocytosis	- $_{1}$ antitrypsin deficiency *
- Cretinism *	- Alagille syndrome *
- Pyloric stenosis	- Zellweger syndrome *

- Drugs: e.g. novobiocin, vitamin K etc.
- Crigler-Najjar syndrome *
- Transient familial hyperbilirubinemia (Lucey-Driscoll syndrome)

(* Prolonged jaundice, i.e. > 10 days in term and > 2 weeks in preterm infants)

Infants of diabetic mother (IDM):

IDM are complications of either established diabetes or of gestational diabetes. In either situation the complications are directly related to the maternal blood glucose level. These infants have a characteristic appearance of a large plump body with a small appearing puffy plethoric face.

IDM are commonly complicated by one or more of the following **problems**:

- *Macrosomia* (large body size)
- *Birth trauma* and/or *intrapartum asphyxia*
- *Hyaline membrane disease*
- *Metabolic disorders* (hypoglycemia, hypocalcaemia, hypomagnesaemia, hyperbilirubinemia)
- *Polycythemia* and its complications (renal vein thrombosis)
- *Visceromegaly* (hepatomegaly, hypertrophic cardiomyopathy)
- *Congenital anomalies* (sacral agenesis, lower limb hypoplasia, small left colon syndrome, ventricular septal defect, transposition of great arteries)
- *Intrauterine growth retardation* (in diabetics with severe vascular disease)

Differential diagnosis of ***large for date*** infants -

- Familial
- Infants of diabetic mother syndrome
- Glycogen storage disease
- Nesidioblastosis
- Hydrops fetalis
- Syndromes
  - Beckwith-Wiedemann
  - Sotos syndrome
  - Marshall-Smith syndrome
  - Weaver-Smith syndrome

Metabolic disorders in newborn:

Owing to the high rate of consanguinity inborn errors of metabolism (IEM) is a relatively common problem in Saudi Arabia and deserves special attention. In any ill looking infant whom presents with either acute or recurrent symptoms, IEM should be kept in mind in addition to other possibilities. Prompt treatment can prevent mental retardation and death. Even in a dying neonate identification of an IEM and subsequent counseling can prevent further catastrophe for the family.

History:

Symptoms of IEM are often non-specific and include poor feeding, vomiting, tachypnea, irritability, altered mental status and seizures. If you are dealing with a neonate who presents with these problems consider the possibility of IEM after excluding other conditions, e.g. infection, CNS pathology, cardiac defects etc. In addition to the points mentioned in pregnancy and neonatal history, emphasis should be given to the following aspects of history -

- Parental consanguinity
- Neonatal deaths in siblings
- Relatives with mental retardation or neurologic disability
- Unusual odor from the body or excretions

(All these point towards metabolic disorders, but a negative family history does not rule out the diagnosis)

Physical examination:
It may not reveal any abnormality. In addition to the scheme mentioned for newborn examination, importance should be given to the following signs, which might give some clue to the diagnosis -

- Facial dysmorphism
- Tachypnea
- Apnea
- Microcephaly
- Macrocephaly
- Hypotonia
- Hypertonia Cataract
- Jaundice
- Hepatomegaly

Investigations:
The mainstay of diagnosis for IEM depends on the laboratory evaluation. Commonly available laboratory tests, e.g. full blood count, blood gases, serum electrolytes and glucose, urine ketones and reducing substances, blood ammonia, lactate and pyruvate can give some clue to the diagnosis and assess the need for further investigations (e.g. amino acids in plasma and urine, organic acids in urine, very long fatty acids in serum etc.)

Newborn metabolic screening:

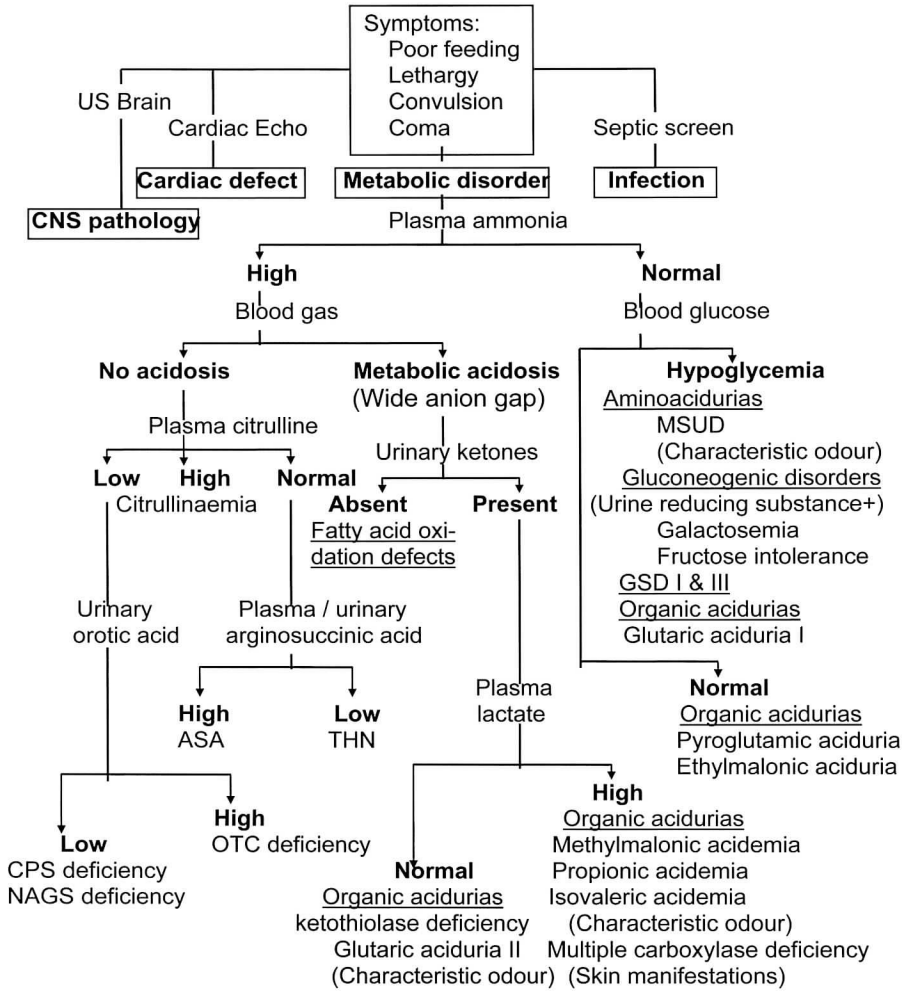
The aim of the screening programme is early detection of those diseases which can be prevented or treated early to avoid their further progression. The selection of the diseases for screening should be guided by their prevalence in the community. It should also be cost effective.

The technique most commonly used for this purpose is the extended *Guthrie test* (blood obtained by heel prick from 3rd day of life and allowed to dry on a filter paper) which is a bacterial inhibition assay developed originally for phenylketonuria, now extended to screen additional metabolic disorders.

An abnormal finding in a newborn screening test is not diagnostic of a disorder; additional tests have to be performed so as to confirm or rule out the condition.

To date *congenital hypothyroidism* is the only nation-wide newborn metabolic screening programme implemented in Saudi Arabia. It is performed on cord blood to measure TSH level. Other screening programmes are underway to be implemented in future.

APPROACH TO METABOLIC DISORDERS IN NEWBORN



Laboratory workup for IEM. Abbreviations: US = Ultrasound sonography, MSUD = Maple syrup urine disease, ASA = Arginosuccinic acidemia, GSD = Glycogen storage disease, OTC = Ornithine transcarbamylase, THN = Transient hyperammonemia of newborn, CPS = Carbamyl phosphate synthetase, NAGS = N-acetyl glutamate synthetase.

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- Ward J C. Inborn Errors of Metabolism of Acute Onset in Infancy. Pediatr in Review 1990; 11:205-216
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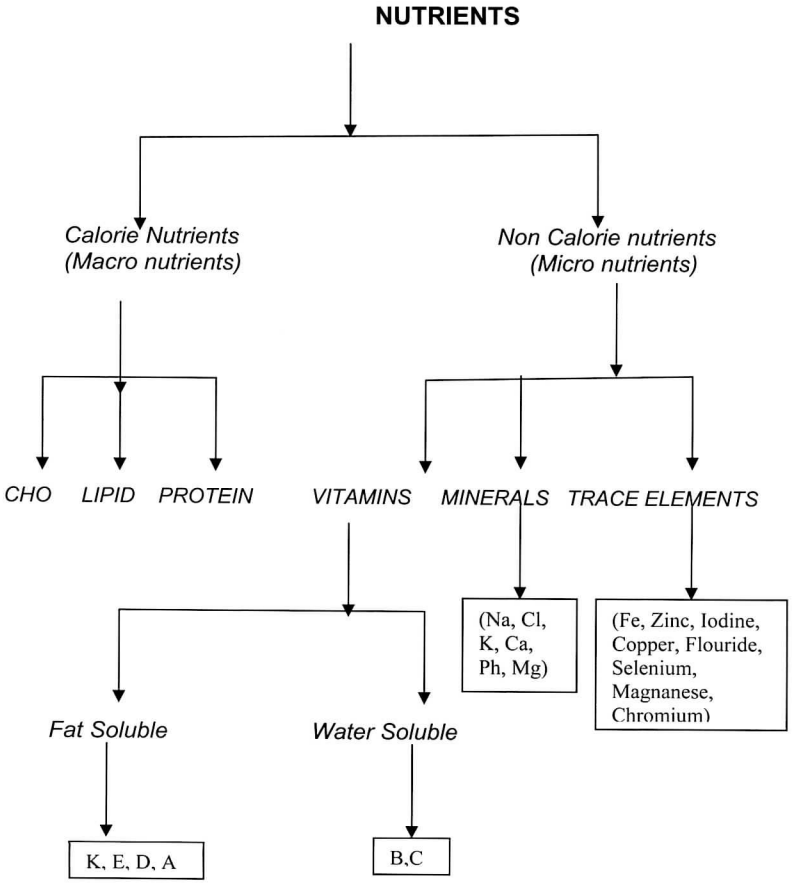
NUTRITION

- **An adequate diet is essential to:**
  1. Maintain body mass
  2. Support activity and play
  3. Allow growth and development.

- **Nutrient adequacy can be evaluated by:**
  1. Dietary history
  2. Examination of growth data and physical examination
  3. Laboratory testing.



NUTRITIONAL REQUIREMENTS:

Category Or condition	Age (years)	Protein (gm)	Vit. A (µg RE*)	Vit. D (µg**)	Vit. E (mg α-TE***)	Vit. K (µg)	Vit. C (mg)	Calcium (mg)	Phosph. (mg)	Magnesium (mg)	Iron (mg)
Infants	0.0-0.5 0.5-1.0	13 14	375 375	7.5 10	3 4	5 10	30 35	400 600	300 500	40 60	6 10
Children	1-3 4-6 7-10	16 24 28	4000 5000 7000	10 10 10	6 7 7	15 20 30	40 45 45	800 800 800	8000 8000 8000	80 120 170	10 10 10
Males	11-14 15-18	45 59	1000 1000	10 10	10 10	45 65	50 60	1200 1200	1200 1200	270 400	12 12
Females	11-14 15-18	46 44	800 800	10 10	8 8	45 55	50 60	1200 1200	1200 1200	280 300	15 15

*Retinol equivalents. 1 retinol equivalent = 1µg retinol or 6 µg β-carotene.
**As cholecalciferol. 10µg cholecalciferol = 400 IU of vitamin D.
*** α - tocopherol equivalents. 1 mg d- α tocopherol – 1 α-TE
Vit. = Vitamin
Phosph. = Phosphorus

RECOMMENDED ENERGY BASIC REQUIRMENT (kcal)

CATEGORY	AGE (YEARS)	PER KG	PER DAY
INFANTS	0.0-0.5	108	650
	0.5-1.0	98	850
CHILDREN	1-3	102	1300
	4-6	90	1800
	7-10	70	2000
MALES	11-14	55	2500
	15-18	45	3000
FEMALE	11-14	47	2200
	15-18	40	2200

CALORIC REQUIRMENT INCREASE IN

FEVER	12% PER 1° C > 37°C
CARDIAC DISEASES	15-25%
MAJOR SURGERY	20-30%
SEVER SEPZIS	40-50%
LONG TERM GROWTH FAILURE	50-100%
BURN	UP TO 100%

Catch up growth requirement for malnourished infants and children.

$$\text{Calories requirements} = \text{estimated requirement} \times \frac{\text{Ideal wt.}}{\text{Actual wt.}}$$

FLUID REQUIREMENT

HOLLIDAY-SEGAR METHOD:	
For the 1st 10 kg body weight	100 ml/kg
For the 2nd 10 kg body weight	50ml/kg
For each additional kg	20 ml/kg
Body Surface Area Method:	1500 ml/m² 24 hrs

NORMAL ELECTROLYTE REQUIREMENTS

ELECTROLYTE	DAILY REQUIREMENT
SODIUM	2-4 mEq/kg
POTASSIUM	2-3 mEq/kg
CHLORIDE	2-3 mEq/kg
MAGNESIUM	0.25-0.5 mEq/kg
CALCIUM	
INFANTS	300-400 mg/kg
CHILDREN	100-200 mg/kg
ADOLESCENTS	50-100 mg/kg
PHOSPHORUS	
INFANTS	1-1.5 mmol/kg
CHILDREN	1 mmol/kg
ADOLESCENTS	0.5-1 mmol/kg

MILLIQUIVALENT – MILLIGRAM CONVERSION TABLE

MINERAL ELEMENT	CHEMICAL SYMBOL	ATOMIC WEIGHT	VALENCE
CALCIUM	Ca	40	2
CHLORINE	Cl	35.4	1
MAGNESIUM	Mg	24.3	2
PHOSPHORUS	P	31	2
POTASSIUM	K	39	1
SODIUM	Na	23	1

$$\text{Milliequivalents} = \frac{\text{Milligrams}}{\text{Atomic weight}} \times \text{valence}$$

* Example – convert 2000 mg sodium to mEq of sodium = $\frac{2000}{23} \times 1 = 87$ mEq sodium

$$\text{Milligrams} = \frac{\text{Milliequivalent}}{\text{Valence}} \times \text{atomic weight}$$

*Example: convert 20 mEq sodium to mg of sodium = $\frac{20 \times 23}{1} = 460$mg sodium

ENTERAL NUTRITION

GENERAL CONSIDERATION:

1. The gastro intestinal tract should be used whenever possible.
2. Small bowel feeding must always be given by continuous infusion.
3. Elemental diets are useful with limited enteric function
4. Continuous enteral drip feeding is better tolerated than bolus feeding when:-
    a. The intestinal mucosal surface has been severely injured.
    b. If there is gastric emptying delay.
    c. During state of hypermobility.

INDICATION:-

Inability to tolerate adequate oral feeding (critically and chronically ill patient).

ADVANTAGE OVER TPN:-

- Safer.
- Easier.
- Less septic and metabolic complication.

ENTERAL TUBE FEEDING ROUTES:-

- Nasogastric.
- Naso-jejonal. } Short term support
-
- Gastrostomy.
- Jejenostomy. } Long term support

NUTRITIONAL SUPPORT GUIDELINES

Currently tolerating adequate diet→ Yes → Continue feeding and ongoing assessment.

↓

No

↓

Pt. is metabolically stable → No → Delay nutritional support.

Correct fluid and electrolyte abnormalities.

↓

Yes

↓

Patient NPO / No bowel sounds →Yes → Begin parenteral nutrition

↓

No

↓

Pt. likely to tolerate std. Formula → Yes →- Begin continuous iso-or hypotonic feeding

↓

- <12 months use formula child was taking at home

- 1-4 years Pediasure or similar formula

↓

- >5 yrs Adult formulas plus multivitamin and supplement for age.

No

↓

Begin modified formula:

- Infants (<1 yr): begin drip feeds at 1 ml/kg*hr. Use medium-chain triglyceride fortified formula, e.g., Pregestimil™, portagen™.
- Older children: may use adult formula such as diluted tolerex™, Osmolyte®, Reabilan™, Isocal™. Initial rate: 1 ml/kg*hr up to a maximum of 20 ml/hr to start

TPN GUIDELINES

A. Calculations

1. Determine the daily fluid requirement.
2. Determine daily caloric requirements

AGE (YR)	KCAL/KG/DAY
0-1	90-120
1-7	75-90
7-12	60-75
12-18	30-60

3. Macronutrients

INITIATION AND ADVANCEMENT OF PARENTERAL NUTRITION			
NUTRIENT	INITIAL	ADVANCEMENT	MAXIMUM
GLUCOSE	5% to 10%	2.5% to 5% day	12.5% peripheral. 18 mg/kg/min (maximum rate of infusion)
PROTIEN	1 g/kg/day	0.5-1 g/kg/day	3 g/kg/day 10%-16% of calories.
FAT	0.5-1 g/kg/day	1 g/kg/day	4 g/kg/day 0.17 g/kg/hr (maximum rate of infusion).

4. ELECTROLYTE AND MNERALS

SODIUM	2-4 mEq/kg/day
POTASIUM	2-3 mEq/kg/day
CHOLRID	2-3 mEq/kg/day
CALCIUM	0.5-2 mEq/kg/day
PHOSPHORUS	0.5-2 mEq/kg/day
MAGNISUM	0.25-0.5 mEq/kg/day

5. VITAMINS AND TRACE ELEMENTS AS PER PHARMACY NORMS

B. Monitoring schedule

LABORATORY STUDIES	INITIALPERIOD	LATER PERIOD
ELECTROLYTES & GLUCOSE	Daily until stable	Weekly
BUN/CREATININE	2 times/wk	Weekly
ALBUMIN OR PREALBUMIN	Weekly	Weekly
Ca^{2+}, Mg^{2+}, P	2 times/wk	Weekly
ALT, AST, ALKP	Weekly	Weekly
TOTAL & DIRECT BILIRUBIN	Weekly	Weekly
CBC	Weekly	Weekly
TRIGLYCERIDES	With each increase	Weekly

INFANT FEEDING

1. BREAST FEEDING:

Breast Feeding is the optimal choice for feeding normal infant.

Advantages:

- Optimal nutritional intake
- Readily available at the proper temperature
- Fresh and free of contamination
- Reduced risk to allergic diseases
- Better mother - infant bonding
- Reduced risk of infection - it contains:
  - Antibodies against certain bacteria and viruses
  - High concentration of secretory IgA
  - Macrophage
  - Lactoferrin

General Considerations in Breast Feeding

- Breast-feeding can be started after delivery as soon as both mother and baby are stable.
- Both breasts should be offered at each feeding for a minimum of 7-10 minutes each and the starting breast should be alternated.
- No supplemental water or formula should be given the first 2 weeks
- Nipples should be allowed to air dry before covering at the end of feeding
- If the infant is satisfied after each nursing period, sleep 2-4 hours and gain weight adequately, the milk supply is sufficient.

Supplements for Breast Fed Infants:

- Vit. K to be administered at birth to all babies (1mg of Vit. K_1).
- Vit. D to be offered at birth for all breast-fed babies (400 1U/day).
- Iron to be offered for all breast-fed babies by 6 months of age.

2. FORMULA FEEDING:

General considerations in bottle feeding

- The bottle should be held such that no air enters the nipple
- Bottle should never be "propped", propping cause choking, caries, and otitis media
- Infant tend to:
  - Swallow air if the nipple hole too small
  - Choke if the nipple hole too large
- When the bottle held upside down the milk should come out in frequent drips (not stream)
- Infant should be burped frequently

Composition of Human Milk and Cow's Milk

	Human Milk	***Cow's Milk***
MACRONUTRIENTS (dl)		
Calories (Kcal)	70	67
Protein (g)	1.1	3.3
Lactose (g)	7	4.8
Fat (g)	3.8	3.8
MINERALS (in liter)		
Calcium (mg)	340	1170
Phosphorus (mg)	150	920
Iron (mg)	0.5	1
Sodium (mEq)	7	25
Potassium (mEq)	14	35
Chloride (mEq)	12	29
VITAMINS (in liter)		
Vitamin A (IU)	1900	1000
Vitamin D (IU)	50	20
Vitamin E (mg)	2.5	0.5
Vitamin K (μg)	15	60
Vitamin C (mg)	50	10

1) MACRONUTRIENTS

- Human milk contains more lactose
- Cow's milk contains more protein

2) MICRONUTRIENTS

A) *MINERALS*

- Cow's milk contains much more of all minerals except iron and copper.

B) VITAMINS

- Human milk contains more vitamins A, C, D and E.
- Cow's milk contains more vitamin K.

INFANT FORMULAS

NAME	Kcal/OZ	CHO	FAT	PROTEIN	INDICATION
Term Formula e.g. Similac, Nan, S-26	20	Lactose	LCT	Cow's milk protein	Term baby Pre term > 2,000 gm
Preterm Formula e.g. Similac Special Care	24	Lactose + glucose polymer	MCT + LCT	Cow's milk protein	Preterm < 2,000 gm
Soy based Formula e.g. Isomil, Nursoy	20	Sucrose + Corn Syrup	LCT	Soy Protein	-Lactase deficiency Primary, secondary -Galactosaemia -Cow milk protein allergy
Lactose free formula e.g. Lactofree	20	Corn Syrup	LCT	Cow's milk protein	-Lactase deficiency 1ry,2ry -Galactosaemia
Fructose free formula e.g. Similac	20	Lactose	LCT	Cow's milk protein	Fructosemia
Glucose free formula e.g. Galactomin	19	Fructose	LCT	Cow's milk protein	Glucose - galactose Malabsorption
Protein Hydrolysate Formula e.g. Nutramigen	20	Corn Syrup	LCT	Protein hydrolysate	-Cow milk protein Allergy -Multiple Food allergy
Fat Modified e.g. Portagen	20	Sucrose + Corn Syrup	MCT + LCT	Cow's milk protein	Steatorrhoea
Elemental formula e.g. Pregestimil	20	Glucose polymer	MCT + LCT	Protein Hydrolysat + A.A.	-Malabsorption -Intestinal resection

LCT = Long chain triglyceride
MCT= Medium chain triglyceride

FORMULAS AVAILABLE FOR SOME METABOLIC DISORDERS

DISEASES	OFFENDING SUBSTANCES TO AVOID	FORMULA AVAILABLE
Maple Syrup Urine Disease	Leucine, Valine, Isoleucine	e.g. Ketonex
Propionic and Methyl malonic acidemia	Valine, Isoleucine	e.g. Propimex
Isovaleric Acidemia	Leucine	e.g. Valex
Phenyl Ketonuria	Phenyl alanine	e.g. phenex
Homocystinuria	Homocystine, Methionine	e.g. Hominex
Urea cycle defect	all amino acid	e.g. cyclinex

WEANING

Definition:
- Is the gradual introduction of food other than breast milk or formula.

Aim:
- To increase the energy density of the diet
- Important source of vitamins and trace minerals.

Onset:
- Weaning should commence not earlier than 4 months, not later than 6 months

Suggested schedule
- First iron fortified infant cereal.
- Next strained fruits and vegetables.
- Progressing to pureed meats and fish.

General considerations in weaning
- Foods are best introduced one at a time.
- Early weaning foods need to be smooth.
- Potentially allergenic items such as egg whites, chocolates are generally introduced after the first birthday.
- Honey is not recommended during the first year of life because may associate with infantile botulism.
- Whole cow milk should not be introduced until after first year of age.

COMMON NUTRITIONAL DISORDERS

I. PROTEIN ENERGY MALNUTRITION

PEM is a spectrum of conditions due to varying proportion of protein and calorie deficiencies.

Causes:

1. Inadequate calorie intake e.g. lack of food, anorexia.
2. Increased calorie loss e.g. malabsorption, cystic fibrosis
3. Increased calorie requirements e.g. infection, trauma.

A) MARASMUS

(Malnutrition is due to severe calorie depletion).

Clinical manifestations:

- Emaciation with body weight below 60% that expected for age.
- Loss of muscle mass and subcutaneous tissue.
- Skin is dry and thin.
- Hair is thin and sparse.
- Weakness and hypotonia.

B) KWASHIORKOR

(Malnutrition is due to inadequate protein intake)

Clinical manifestations:

- Edema with body weight ranges from 60-80% of the expected for age.
- Loss of muscle mass
- Skin changes are common and range from hyperpigmented hyperkeratosis to an erythematous macular rash.
- Hair is sparse, easily pluckable and appears dull brown, adequate protein intake restores hair color (Flag sign).
- Irritability and secondary immunodeficiency.

Treatment of Protein energy malnutrition

- Acute management of shock, infection and organ failure.
- Fluid and electrolyte replacement
- Nutritional support
    1. Start with 50-75% of the normal energy requirement
    2. Calorie intake can be increased 10-20%/day
    3. Calorie intake is increased until appropriate regrowth is initiated this may require 150% or more of the recommended calories.
    4. Protein needs are provided in proportion (10-20%) to the calorie intake.
    5. Vitamin and mineral intake in excess of recommended daily allowance is provided.

2. OBESITY

(Obesity is due to excess intake of calories)

Clinical manifestation

- Heavy (actual weight greater than 120% of ideal weight for age, sex and height.
- Tall
- Advanced bone age.

Treatment

- Education
- Behavior modification
- Exercise
- Drug (usually reserved for severely obese adolescent patients)
- Diet - consisting of an approximately 30% decrease in previous calorie intake.

3. NUTRITIONAL RICKETS

(Failure of mineralization of growing bone or osteoid tissue due to Vitamin D deficiency)

Clinical manifestations

- Thickening of the wrist and ankles
- Anterior fontanel is enlarged
- Craniotabes
- Enlargement of costochondral junction (rachitic rosary)
- Scoliosis
- Bow legs

Treatment: Vitamin D 4000 - 5000 IU/day

4. IRON DEFICIENCY ANEMIA

(The major causes are a decrease of body iron by rapid growth and an iron poor diet)

Clinical manifestations

- Pallor
- Spoon nail
- Reduce muscle and mental performance

Treatment: Iron as ferrous sulfate 6 mg/kg/day

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IMMUNIZATION

IMMUNITY:
The body can acquire protection through

1. Passive immunity:
   The protection is immediate but for a brief duration. It is acquired through
   a) Natural - Maternal Antibodies (through placenta)
   b) Artificial - Immunoglobulin or Antitoxins
2. Active immunity:
   Here immunity can be acquired when the antigen is introduced to the host (human body), which produces its own antibodies. This type of protection takes 1-2 weeks to develop but is of longer duration. It occurs through
   a) Natural – Infection
   b) Artificial – Immunization with vaccines

Vaccines could be divided into inactivated (killed) and live attenuated vaccines. The characteristics of each are discussed in the following table:

STATUS OF VACCINE ANTIGEN

	LIVE	KILLED
VIRULENCE	Attenuated	Noninfectious
Replication	Yes	No
Effect of passive immunity	May prevent successful immunization	No inhibition effect
Duration of immunity	Longer	Shorter
Booster	Usually no need	Yes
Adverse reaction	Can produce disease especially in immuno-compromised children	Low incidence of adverse reaction
Examples		
Whole-cell bacteria	BCG	Pertussis
Whole-cell virus	Measles Mumps Rubella Oral Polio (OPV)	Injectable polio
Toxoid		*Tetanus*
Polysaccharide		Pneumococcal
Recombinant protein		Hepatitis B

Basic Vaccination schedule in K.S.A. 2009

Age	Vaccine
At Birth	BCG, Hepatitis B
2 months	IPV, DTP, Hib, PCV, Hepatitis B
4 months	OPV, DTP, Hib, PCV,Hepatitis B
6 months	OPV, DTP, Hib, Hepatitis B
9 months	Measles (mono)
12 months	OPV, MMR, Varicella, PCV,
18 months	OPV, DTP, Hib, Hepatitis A
24 months	Hepatitis A
4-6 years	OPV, DTP (DT) MMR Varicella

Suggested vaccination schedule for young children with no previous vaccination and above one year of age

VISIT	Interval From the previous visit	VACCINE
1ST	-----	IPV [DPT+HIB + + HBV] MMR, Varicella , PCV
2ND	2 months	OPV [DPT+HIB + + HBV]
3RD	2 months	OPV [DPT+HIB + + HBV]
4TH	2 months	BCG
5TH	6-12 months	OPV [DPT+HIB + + HBV] Hepatitis A
6th	6 months	Hepatitis A
7TH	4-6 years	OPV + DPT MMR, Varicella

GOLDEN RULE

Simultaneous administration of vaccine in the same visit produces the same immunogenic response as the single vaccine.

GENERAL CONTRAINDICATIONS TO IMMUNIZATION

- Fever associated with moderate or severe illness or in an immunocompromised child: postpone immunization.
- Hypersensitivity reaction to the vaccine or its constituents.

CONTRAINDICATION TO LIVE ATTENUATED VACCINES

- Patients with immunodeficiency; congenital immunodeficiency, leukemia, lymphoma, solid tumors, or those with HIV infection.
  (An exception is measles, MMR vaccination, which is recommended for both asymptomatic and symptomatic HIV - infected children).
- Those being treated with immunosuppressive medications or large or prolonged (>14 days) doses of systemic corticosteroids.
- Within 3 weeks of another live vaccine; live vaccines should be given simultaneously or at least 4weeks apart.
- Recent administration of immunoglobulin (within 3-11 months depending on the product and the dose)
- Patient with tuberculosis should not receive measles vaccine unless on full treatment for TB

False or Invalid Contraindication to Immunization

- Mild upper respiratory infection
- Treatment with antibiotics
- Prematurity and low birth weight
- Older than the usual age for immunization
- Breast feeding, mild diarrhoea (oral polio vaccine)
- Contact with an infectious disease
- Malnutrition
- Short courses (< 2 weeks), or physiological maintenance doses of corticosteroids
- History of nonspecific allergies.

VACCINATION IN SPECIAL CIRCUMSTANCES

Children whose immunization status is unknown:

- No harm to vaccinate immune children.

Children whose immunization schedules were not completed:

- Continue and do not start from the beginning.

Preterm infants:

- Vaccinate according to chronological and not gestational age. (Do not reduce or divide the dose.)
- At time of birth, may delay hepatitis vaccine if birth weight < 2 kg and mother is HBsAg negative. If positive, do not count birth dose.
- Consider influenza vaccine in infants with bronchopulmonary dysplasia.

Children with congenital or acquired immunodeficiency:

- Do not give live attenuated vaccines. Under certain conditions, BCG, MMR may be given to children with asymptomatic HIV infection; Infectious diseases consultation is needed.
- Injectable polio vaccine (IPV) should be given instead of oral polio to the infants and their contacts.

Hospitalized patients:

- Do not give OPV during hospitalization. OPV can be given on discharge, or if prolonged hospitalization is expected, give IPV.

Asplenic or splenectomized children:

- Pneumococcal, HIB and meningococcal vaccine should be given in addition to routine vaccination.
- When elective splenectomy is performed, give vaccines 2 weeks or more before the operation.

ACTIVE IMMUNIZATION AFTER EXPOSURE

In the following diseases vaccination can protect children after exposure to the disease, if it is given early. Incubation period is longer than the time needed to induce immunity by vaccination.

- Measles - within 72 hours of exposure
- Hepatitis B - as soon as possible & accompanied by hepatitis B immunoglobulin
- Hepatitis A - as soon as possible and within 2 weeks of exposure.
- Tetanus - depends on vaccination history, also needs passive immunization with specific immunoglobulin depending on the nature of the wound and vaccination history.
- Rabies - part of the treatment to induce immunity.
- Varicella - within 72 hr of exposure, but up to 5 days is acceptable.

B C G VACCINE

Nature of the vaccine:

- The BCG vaccine is live attenuated vaccine containing the attenuated TB bacilli, *mycobacterium bovis*.

Properties:

- It gives reasonable protective efficacy against systemic tuberculosis (miliary, meningitis) [up to 75%] and variable efficacy against pulmonary tuberculosis.

Administration of the vaccine:

- Single intradermal injection in the skin on the lateral aspect of the left upper arm overlying the insertion of deltoid muscle.
- Dose: 0.05ml for infants < 12 months, 0.1ml for children > 12 months and adults

Indications:

- Primary vaccination to all newborns
- Infants above 2 months, give only to *PPD negative* individuals
- In children with immunodeficiencies (congenital or acquired), the WHO recommends that in areas with high prevalence of TB, BCG should be given to all infants at birth regardless of maternal HIV infection, if the infants are asymptomatic.

Adverse reaction:

- A superficial, self-healing ulceration may develop after intradermal injection of the vaccine. If oozing occurs, only dry dressings should be used temporarily.
- Inadvertent injection in the subcutaneous tissue may lead to formation of an abscess at the injection site.
- Regional lymphadenopathy.
- Disseminated disease (rare) may occur if the vaccine is given to the child with immunodeficiency.

Note:

- BCG vaccine does not provide absolute protection against TB.
- TB should be considered as a possible diagnosis in any patient who presents with a suggestive history, or signs or symptoms of TB, regardless of immunization history

DIPHTERIA, TETANUS & PERTUSSIS (DTP, DTaP)

Nature of the vaccine:

- The DTP vaccine is a combination of diphtheria and tetanus toxoid with whole cell killed pertussis.
- Acellular pertussis (aP) is available in combination with diphtheria and tetanus toxoids, and contains several antigens from the pertussis organism but with minimal endotoxin.

Preparations:

- < 7 years: DTP, DT, and DTaP
- >7 years: Td, Tdap.
- Preparations with a small 'd' contain the reduced dose of diphtheria toxoid and are for booster doses only and not for primary vaccination.
- There are many commercial preparations that combine the above with *Haemophilus influenzae* type b vaccine and/or inactivated polio vaccine.

Site and method of vaccination:

- IM, in the anterolateral aspect of the thigh.

Indications:

- Primary vaccination to all infants at 2, 4, 6 months.
- Booster vaccination at 18 months and 4-6 years.

Contraindication:

- An immediate anaphylactic reaction to vaccine or vaccine component
- Encephalopathy within 7 days (severe, acute, central nervous system disorder, unexplained by another cause).

Precautions:

The following precautions were considered before as contraindication (especially with the whole cell vaccine. For the acellular vaccine these are not contraindications)

- Convulsion with or without fever within 3 days of DTP/DTaP
- Persistent, inconsolable crying for 3 or more hours within 48 hours
- Collapse or shock-like syndrome within 48 hours
- Temperature 40.5°C or more

Decision to vaccinate should be considered on an individual basis

Reasons for deferral of pertussis vaccine:

- Progressive neurologic disorder
  e.g. infantile spasm, progressive encephalopathy
- Personal history of convulsion
- Infants with condition known to predispose to convulsion
  e.g. tuberous sclerosis
  N.B. Each visit the child should be evaluated and vaccine considered on an individual basis

Adverse reactions

- Local: redness, swelling, and pain
- General: fever, drowsiness, anorexia, vomiting, persistent crying, convulsions, and collapse with shock – like syndrome

NB:

- The acellular vaccine has lower incidence of local and general adverse reactions compared to whole cell and, when available, is the preferred agent for immunization. Also, it can be used for vaccination in adolescents.
- To minimize febrile response, give acetaminophen just before the injection and then q4h x 24hr.

HAEMOPHILUS INFLUENZAE TYPE B (HIB)

Nature of vaccine:

- Polysaccharide vaccine conjugated to a carrier protein e.g.: mutant diphtheria toxin (HIBTITER), tetanus toxoid (ActHIB), etc. They can be combined with other vaccines e.g. DTP.

Properties:

- To ensure > 90% efficacy the number of doses of the vaccine depends on the age of starting the vaccine
- The vaccine can prevent not only *Haemophilus influenzae* meningitis but also epiglottitis, cellulitis and osteomyelitis caused by *Haemophilus Influenzae* type B.

Preparations:

- Various preparations are available; either alone e.g. ActHB or combined with DTP, IPV

Site and method of vaccination:

- IM, Anterolateral aspect of the thigh

AGE OF STARTING THE VACCINE	NO. OF DOSES
1 - 6 Months	3 + Booster at 15 months
7 -11 Months	2 + Booster
11-14 Months	1 + Booster
15 Months	1

Indications:

- Primary vaccination to all infants at 2, 4, 6 months

Contraindication:

- An anaphylactic reaction to vaccine or vaccine component

Adverse reaction:

- General: Minimal
- Local: Mild pain, swelling

HEPATITIS A VACCINE

Nature of vaccine:

- Killed vaccine, formalin inactivated viral particles.

Preparations:

- Various monovalent preparations are available e.g. Havrix Junior® [720 ELISA units] Avaxim, and Vaqta Paediatric® for children, Havrix® [1440 ELISA units] for adults etc.
- Also available in combination with Hepatitis B vaccine e.g. Twinrix®

Indications:

Children 12 months and above

- *Primary immunization*
- *Susceptible children in endemic areas*
- *Chronic liver disease*
- *Hemophilia*
- *Post exposure prophylaxis to susceptible contacts of hepatitis A cases: alone or in combination with human immunoglobulin*
    - If IG is considered, give within 2 weeks of exposure

Site and methods of vaccination:

- IM (SC in hemophilic children)
- 2 doses, at least 6 months apart
- If given with immunoglobulin, give at a different site

Contraindication:

- Hypersensitivity to vaccine or vaccine component.

Adverse Reaction:

- Local reactions, fever
- Rare: anaphylaxis

HEPATITIS B VACCINE

Nature of the vaccine:

- The vaccine contains purified HBsAg produced from yeast cells using recombinant DNA technology. The vaccine contains inactivated viral particles with no live organisms.

Properties:

- The vaccine protects against the hepatitis B virus by producing specific antibodies to HBsAg (anti-HBs). Immunization against hepatitis B also affords indirect protection against the delta virus, a defective virus that can replicate only in subjects infected by the HB virus

Preparations:

- Engerix®-B
- Recombivax HB®
- Combined with Hepatitis A vaccine: Twinrix®, & Twinrix® Junior

Site and method of vaccination:

- IM in the outer part of the thigh in infants or deltoid muscle in older children.

Note:

- Children (0-15 years) receive ½ the adult dose.
- Children on haemodialysis need double the adult dose.

Indications:

- Primary vaccination to all newborns.
- *Pre-exposure prophylaxis for*
    - Health care and emergency service workers
    - High risk groups such as: injection drug users, hemophiliacs and patients receiving repeated infusions of blood or blood products, hemodialysis patients, those with illicit sexual contacts, and

household or sexual contacts of acute HBV cases and HBV carriers, , etc.

- *Post-exposure prophylaxis* (consider also HBIG) for:
  - Infants born to infected mothers
    - Infants born to HBsAg positive mother should receive immunoprophylaxis (HBIG) and the first injection of the vaccine after birth in two different sites.
    - In premature newborn < 2 kg. The dose given at birth is not counted. The infant should receive should receive 3 further doses after it is > 2kg. or > 2 months.
  - Percutaneous (needle-stick, bite) or mucosal exposure
  - Sexual and household contacts of hepatitis B:

Contraindication:

- Allergy to any component of the vaccine
- Previous anaphylactic reaction

Adverse reaction:

- Local reactions: Pain, induration (1- 6 % of cases)
- Systemic reactions: Fever, muscle pain (< 1% of cases)

INFLUENZA VIRUS VACCINE

Nature of vaccine:

- Both inactivated and live attenuated vaccines (LAIV) are available.
- They include 3 strains, 2 type A and 1 type B
- The composition of the vaccine is changed annually.

Preparations:

- Inactivated influenza vaccine: e.g. Flusone for children above 6 months
- Live attenuated influenza vaccine (LAIV): Flumist only for healthy children above 2 years with no underlying medical problem

Indications for Inactivated influenza vaccine:

- Patient with chronic cardio-respiratory disease (e.g., asthma, cystic fibrosis, bronchopulmonary dysplasia)
- Sickle cell anemia
- Those with chronic salicylate therapy
- Those with chronic conditions: Diabetes mellitus, Chronic renal disease, Chronic metabolic disease
- Patients with immunosuppressive conditions: cancer, HIV etc.
- Hospital personnel with significant patient contact

Site and methods of vaccination:

- Inactivated vaccine = IM, LAIV = intranasal; divide dose among each nostril
- 1 dose during influenza season
- Children younger than 9 years and not previously immunized should receive an additional dose, 4 weeks after the 1st dose for the first 2 years.

Contraindication:

- Inactivated IV:
    - Anaphylaxis to previous dose
    - Hypersensitivity to eggs
- LAIV: same +
    - immune deficiency, asthma, history of Guillan-Barre Syndrome

Adverse Reaction:

- Soreness at injection site, fever, myalgia
- Allergic response,
- Guillan-Barre Syndrome

MEASLES, MUMPS & RUBELLA (MMR)

Nature of the vaccine:

- Contain the live attenuated viruses of measles, mumps and rubella

Preparations:

- Monovalent measles vaccine
- Combined: MMR II , Priorix®

Site and methods of vaccination:

- SC Injection (anterolateral aspect of the thigh)

Indications:

- Primary vaccination
    - Monovalent measles vaccine: at 9 months
    - MMR, 2 doses, at 1 and 4-6 years

Adverse Reaction:

The combined vaccination is well tolerated in children. Reactions that may be observed from the 5th day after the injection include:

- Fever, Mild rash
- Joint manifestations
- ITP within 6 weeks (controversial, uncommon)
- Parotitis, orchitis (rare)
- Neurological complications such as febrile convulsions are rare.

Contraindications:

- See under contra indications to live attenuated vaccines
- Anaphylactic reactions to neomycin or a previous dose of the vaccine.
- Whether an anaphylactic reaction to eggs is also a contraindication is controversial. Some advise vaccination under hospital controlled conditions
- ITP within 6 weeks. (controversial)
  Note: If PPD skin testing for tuberculosis is required, it should be done on the same day as immunization or delayed for 4-6 weeks.

MENINGOCOCCAL VACCINE

Nature of vaccine:

- 2 types are available
  - Purified capsular polysaccharide vaccine &
  - Protein-polysaccharide conjugate vaccine
- No vaccines are available against serogroup B

Preparations:

- Polysaccharide vaccine (above 2 years);
  - Quadrivalent polysaccharide A/C/Y/W 135(avalable in Saudi Arabia
- Protein conjugate vaccine (For children 2 months and above)
  - Meningococcal group C vaccine
  - Quadrivalent conjugale vaccine A/C/Y/W 135

Indications:

- Polysaccharide vaccine
  - During epidemics caused by Meningococcal A/C/Y/W 135
  - Travel to an endemic area e.g., Mecca during pilgrimage (Hajj)
  - Sickle cell anemia
  - Functional or anatomic asplenia
  - Patient with terminal complement deficiency
- Conjugate vaccine
  - For primary immunization for infants in UK and Canada
  - For primary immunization for adolescents 11-18years in USA.

Site and methods of vaccination:

- Polysaccharide vaccine: SC
  - 3-23 months: 2 doses 2 weeks apart; booster after 1-2 years (only if protection against serotype A is needed)
  - 2-5 years: 1 dose; booster after 2-3 years
  - 5 years and above: 1 dose; booster after 5 years or more
- Protein conjugate vaccine: IM
  - For primary immunization for infants < 1 year in UK and Canada: 3 doses at least 1 month apart
  - For primary immunization for adolescents 11-18years in USA: 1 dose

Contraindication:

- Hypersensitivity to vaccine or vaccine component.
- Conjugate vaccine: history of Guillan-Barre Syndrome

Adverse Reaction:

- Mild reactions; pain & local redness, fever
- Rare: anaphylaxis
- Conjugate vaccine: ? Guillan-Barre Syndrome, transverse myelitis

PNEUMOCOCCAL VACCINE

Nature of vaccine:

- Pneumococcal polysaccharide vaccine (PPV) contains purified capsular polysaccharide from each of 23 capsular types of pneumococcus
- Pneumococcal conjugate vaccine (PCV) contains polysaccharide who are conjugated to protein (CRM197)

Preparations:

- 23 Strain Polysaccharide, for children 2 years and older. (Pneumovax®)
- 7-valent conjugated vaccine for children < 2 yr (Prevnar®)

Indications:

Polysaccharide vaccine

- Sickle cell anemia
- Functional or anatomic asplenia
- Nephrotic syndrome
- Chronic pulmonary disease
- Primary immunodeficiency, Immunosuppression
- CSF leaks
- Cochlear implants

Conjugate vaccine

- Routine vaccination for children < 23 months in selected countries

Site and methods of vaccination:

- Polysaccharide vaccine: IM
- Conjugate vaccine: IM, at 2, 4, 6 & booster at 12-15 months of age. If 1-2 years: give only 2 doses 2 months apart. If 2-5 years: 1 dose.

Contraindication:

- Anaphylaxis to vaccine or vaccine component.

Adverse Reaction:

- Common: local erythema, soreness, slight fever.

POLIOMYELITIS VACCINES

Nature of vaccine:

- Oral Polio Virus Vaccine (OPV) contains attenuated poliovirus type 1, 2 and 3
- Inactivated Polio Virus"Salk" (eIPV) formalin inactivated poliovirus type 1, 2 and 3 with enhanced potency

Preparations:

- Oral Polio Virus Vaccine (OPV)
- Inactivated Polio Virus "Salk" (eIPV) formalin inactivated poliovirus type 1, 2 and 3 with enhanced potency

Indications:

- Primary vaccination of infants

- IPV is the vaccine of choice in immunodeficient children and to people who will have household or close contact with immunodeficient children

Method of vaccination:

- OPV: Oral (behind the tongue)
- IPV: SC injection, IM when part of a combination vaccine.

Contraindications:

- OPV See under "contraindication to live attenuated vaccine."
- IPV: Anaphylaxis to vaccine or vaccine component

Precautions:

- OPV
  - Vomiting of the dose within 5-10 minutes requires giving the dose again
  - If there is persistent vomiting and, or severe diarrhea, postpone the vaccination till the conditions subside. The WHO recommends not to withhold OPV during diarrhea, but to give an extra dose 4 - 6 weeks later.

Adverse reaction:

- OPV: Paralysis due to reversion of viral attenuation can be observed in vaccinated persons or unimmunized contacts (within 60 days). Rare (fewer than 1 in 1 million doses)
- IPV: minor local reactions, anaphylaxis (rare)

ROTA VIRUS VACCINE

Nature of the vaccine:

- RotaTeq contains 5 reassortant rotaviruses developed from human and bovine (WC3) parent rotavirus strains
- Rotarix live monovalent oral vaccine originating from a G1P[8] strain that was isolated from a case of infantile gastroenteritis.

Preparations:

- RotaTeq
- Rotarix

Site and method of vaccination:

- Rotarix vaccine: PO at 2 and 4 months of age.
  - 1st dose: can be given at the age of 6 weeks and should be given no later than at the age of 12 weeks.
  - The interval between the 2 doses should be at least 4 weeks.
  - The 2-dose schedule should be completed by age 16 weeks, and no later than by 24 weeks of age.
- RotaTeq vaccine: PO at ages 2, 4 and 6 months.
  - The first dose should be given between ages 6–12 weeks
  - Subsequent doses at intervals of 4–10 weeks.
  - Vaccination should not be initiated for infants aged >12 weeks.

- All 3 doses should be given before the age of 32 weeks.

Indications:

- Potential use as primary vaccination in young infants

Contraindication:

- Hypersensitivity to any of their components
- History of intussusception or intestinal malformations
- Vaccination should be postponed in case of ongoing acute gastroenteritis or serious febrile illness
- There is a potentially higher risk of intussusception when the fi rst dose of these vaccines is given to infants aged >12 weeks

Adverse reaction:

- mild and transient symptoms from the gastrointestinal or respiratory tract
- avoid vaccinating infants who are potentially immunocompromised, including HIV infected individuals, and to infants with pre-existing chronic gastrointestinal till further information is available

Note

WHO strongly recommends the inclusion of rotavirus vaccination into the national immunization programs where

- vaccine efficacy data suggest a significant public health impact
- appropriate infrastructure and financing mechanisms are available for vaccine use

To date, the clinical efficacy of rotavirus vaccines has been demonstrated mainly in the USA, Europe and Latin America. The WHO awaits data on vaccine efficacy from clinical trials that are currently ongoing in Africa and Asia, where rotavirus disease burden is very high before recommending global inclusion of rotavirus vaccines into national immunization programs.

VARICELLA VACCINE

Nature of vaccine:

- Lyophilized, live, attenuated varicella virus

Preparations:

- Varivax® III
- Varilrix®
- Varicella vaccine is also combined with MMR as MMRV vaccine

Indications:

- Primary vaccination of all susceptible children 12 months and above
- Selected groups with immunodeficiency may be vaccinated under guidance of infectious diseases consultant with vaccination expertise.

Site and methods of vaccination:

- 12 months and above: 2 doses SC at least 4 weeks apart
- Given as primary vaccination at 1 & 4-6 years of age

Contraindication:

- Anaphylaxis to vaccine or vaccine component.
- Children with T-cell immunodeficiency.

Adverse Reaction:

- Mild local reactions as pain, swelling and redness at injection site
- Low grade fever
- Varicella-like rash

TYPHOID VACCINE

Nature of vaccine:

- Oral: vaccine contains live attenuated strain *S. typhi* Ty21a,
- Parental: capsular polysaccharide vaccine of *S. typhi* strain TY2.

Preparations:

- *Parenteral, capsular polysaccharide vaccines (Typh-I)*
  - Typherix®
  - Typhim Vi®,
- *Oral, live attenuated vaccines (Typh-O)*
  - Vivotil®,
  - Vivotif L®,

Indications:

- Food handlers
- Laboratory personnel
- Household contacts.

Site and methods of vaccination:

- Vi capsular polysaccharide vaccine (IM)
  - 2 years and above: Single dose, IM; booster q 3 years
- Oral
  - 3-5 years: 1 sachet, reconstituted, give alternate days x 3 doses
  - >5 years: 1 capsule, alternate days, x 4 doses.

Contraindication:

- Parental: severe local or systemic reaction to a previous dose of the vaccine
- Oral: Acute gastroenteritis, Inflammatory bowel disease, immunpcomprmised individuals

Adverse Reaction:

- Parental: local reactions, fever
- Oral: vomiting, diarrhea, reactive arthritis (rare)

SUMMARY OF COMMONLY USED VACCINES

VACCINE	*TYPE*	ROUTE	STORAGE
BCG	Live attenuated bacteria	ID	2-8°
DTP	Toxoid (D&T) Inactivated bacteria (P)	IM	2-8°
Oral Polio **Inj. Polio**	Live attenuated virus Inactivated virus	Oral IM	2-8° 0-8°*
Hepatitis B	Inactivated virus	IM	2-8°
Measles	Live attenuated virus	SC	2-8°
MMR	Live attenuated virus	SC	2-8° use within 8 hr
HIB conjugate	Polysaccharide-protein conjugate	IM	2-8°
Pneumococcal	Polysaccharide Conjugate	SC IM	2-8°
Meningococcal	Polysaccharide Conjugate	SC IM	2-8°
Influenza	Inactivated virus Live attenuated	IM Intranasal	2-8°
Varicella	Live attenuated virus	SC	2-8° use within 30 min

* Stored at 0-8^{0}C in immunization center but should be consumed within 30 days
N.B. no vaccine should be kept in the freezer

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INFECTIOUS DISEASES

MENINGITIS

- ***Definition***:
  Meningitis is defined as CNS infection
- ***Etiology***
  1. Viral (most common)
  2. Bacterial
  3. Others
- ***Clinical manifestation:*** according to patient's age but in general:
  - Non specific: fever, anorexia, myalgia, arthralgia and various cutaneous signs such as petechia, purpura or erythematous macular rash,
  - Specific (signs of meningeal irritation): Nuchal rigidity, back pain, Kernig sign, Brudzinski sign
  - Symptoms and sign of increased intracranial pressure
- ***Diagnosis:***

Lumber puncture (L.P.): CSF analysis is the cardinal test for diagnosis of meningitis. CSF examination should be done for the followings: Gram stain, Latex agglutination test, Glucose, Protein, Total and differential WBCs, CSF culture and sensitivity. Other tests also indicated: CBCs, Serum glucose, Urea and electrolytes, Blood culture. Other tests can be done for the CSF if indicated: Viral titer or antigen or culture, PCR, Acid fast bacilli, culture for tuberculosis.

CSF findings in different types of meningitis:

CSF	Normal	Viral	Bacterial
Glucose	60% of blood glucose	Normal	Low
Protein	10-40 mg/dL	Usually normal	Usually high
WBCs	Up to 5 lymphocytes	< 1500/cumm	Above normal
Gram Stain	No bacteria	Negative	±
Latex test	Negative	Negative	±
CSF culture	Negative	Negative	±

Contraindications of lumber puncture:

1. Evidence of increased intracranial pressure
2. Severe cardiopulmonary compromise
3. Infection in the skin overlying the site of L.P.
4. Thrombocytopenia (relative contraindication)

- ***Treatment protocol for bacterial meningitis***

Patients' age	Possible organisms	Treatment
Infant < 2 months	E. Coli, Group B streptococcus, Listeria monocytogenes, Salmonella	Cefotaxime + Ampicillin
>2 month	Haemophilus influenza, Pneumococcus	Ceftriaxone or Cefotaxime
Immunocompromised and/or gram negative bacteria		Ceftazidime + Aminoglycoside

- Empirical antibiotic therapy:

- Dexamethasone is indicated prior to antibiotics in all patients except:
  1. Neonate
  2. Patients already on oral or IV antibiotics prior to diagnosis
  3. Pneumococcal meningitis
- Empirical antibiotics should be evaluated after result of CSF culture become available.
- Ceftriaxone can be gien either in single daily dose or divided 12 hourly.
- If gram stain or latex revealed pneumococcus:
  - Stop dexamethosone as it will impair Vancomycine penetration through the blood brain barrier.
  - Vancomycine should be added to Ceftriaxone from the start till the sensitivity of the organism is known.
- Duration of treatment in uncomplicated cases:

N. Meningitides	7 days
H. Influenza	10 days
Strepto. Pneumonia	14 days
Other gram –ve bacteria	3 weeks

Patient's age as well should be considered in deciding the duration of treatment (neonate need longer duration of therapy).

- All patients with meningitis should be kept in isolation for at least 24 hours of starting treatment.
- All patients with meningitis should be reported to health authority.
- Hearing assessment should be arranged before discharge for all patients.
- Long term follow up (at least 6 months) should be arranged.

- Chemoprophylaxis:

- *Haemophilus influenzae type b:*
  - If any of the household contacts is < 4 year old and not fully immunized for Haemophilus influenzae type b, Rifampicin prophylaxis should be given for all household contacts (including the patient if he is not treated with ceftriaxone) immediately after diagnosis is confirmed.
  - Rifampicin 600 mg/dose once daily x 4 days (for adult)
  - Rifampicin 20 mg/kg/dose once daily x 4 days (for children)
- *N. meningitides:*
  - Rifampicin prophylaxis should b given to all household contacts regardless of age as soon as possible.
  - Rifampicin 10 mg/kg/dose twice daily for 2 days (for children), 300 mg/dose twice daily for 2 days (for adults).
  - Health care workers whom in close contact with the patient need to receive Rifampicin as prophylaxis (Ciprofloxacillin can be used as an alternative to Rifampicin, the dose is 500 mg as a single dose only).
  - N.B.: Daily dose of Rifampicin should not exceed 600 mg.
  - Pregnant household contact can be given single dose of intramuscular Ceftriaxone injection (250 mg)
- *Strepto. Pneumonia: no prophylaxis is required*

BRUCELLOSIS

- Clinical manifestation

Fever and sweats
Anorexia, weight loss, weakness, malaise, arthralgia, myalgia, abdominal pain, headache
Lymphadenopathy, hepatosplenomegaly and arthritis

- Diagnosis:

Definitive:

Positive Culture: Blood, Bone marrow or other tissues.

Presumptive:

Serum agglutination test (SAT):

- Positive if titer is 1:160 or greater.
- Single titer is not diagnostic.
- **Early infection will give low titer.**
- Demonstrating raising titer is essential.
- IgM agglutinins can persist in the serum for months or even years, so cannot be used as an index for response to treatment.
- Elevated IgG titer can be found in acute, chronic or relapse.
- Give false positive result (Gram-negative bacteria).
- Prozone phenomena: false-negative results in presence of high antibody titer.
  R, dilute serum to 1:320 or higher as routine.

Enzyme immunoassay (EIA):

- Most sensitive method for determining IgG, IgA and IgM antibrucella antibodies.

Treatment protocol for brucellosis:

	> 8 years old	< 8 years old
Simple infections	• 1st drug: Doxycycline/Tetracycline (6 weeks) • 2nd drug - Bactrim (3 weeks) or - Rifampicin (6 weeks)	○ 1st drug: Bactrim 6 weeks ○ 2nd drug: Rifampicin 6 weeks
*Serious infection or complications: - Endocarditis - Osteomyelitis - Meningitis	- Use Streptomycin (2 weeks) or Gentamicin (1-2 weeks) in place of the 2nd drug. N.B. Rifampicin can be used as well as adjunctive therapy to reduce the rate of relapse. - For life threatening complication such as meningitis or endocarditis, the duration of therapy is often extended for several months. - For osteomyelitis, early surgical intervention should be considered.	

DRUG	DOSE/DAY	Divided to
Bactrim	Trim. 10mg/kg/day + Sulph. 50mg/kg/day	2 doses
Rifampicin	20 mg/kg/day	1 or 2 doses
Doxycycline	2-4 mg/kg/day	2 doses
Tetracycline	30-40 mg/kg/day	4 doses
Streptomycine	20 mg/kg/day (IM)	2 doses
Gentamicin	5 mg/kg/day	3 doses

NB. 1st drug should be given to all patients
Inform infection control department in all positive cases

ENTERIC FEVER

- Clinical manifestation:

Fever
Constitutional symptoms: e.g. Headache, malaise, anorexia and lethargy
Abdominal pain, tenderness, splenomegaly and hepatomegaly
Rose spot (maculopapular rash)
Change in mental status

- Diagnosis:

1) Cultures: Blood culture
Urine and stool culture after the first week
Bone marrow culture (the most sensitive method)
2) Serology: Widal test

- Treatment:

Empirical therapy with Ceftriaxone or Cefotaxime until antibiotic susceptibility is available
In case of resistance to Ceftriaxone, Ciprofloxacillin can be used as alternative.

TUBERCULOSIS

- Clinical manifestation:

General: Fever, night sweat, anorexia, decreased activity, weight loss…
Local: According to site involved

- Diagnosis:

- High index of suspicion
- High ESR
- Positive PPD test (Mantoux test)
- Chest x ray
- Positive smear for acid-fast bacilli and/or culture for mycobacterium tuberculosis from sputum, gastric aspirate, lymph node or other involved site.
- Mantoux test: is intradermal injection of 0.1 ml containing 5 tuberculin units (TU) of purified protein derivative (PPD)
- Interpretation of PPD skin test: PPD considered positive if the amount of indurations after 48-72 hours is either:

5-9 mm with either:	**10-14 mm with either:**	**≥ 15 mm**
1- History of contact with open case 2- Suspected tuberculosis (symptoms, signs or chest radiograph) 3- Immunocompromised patient	- Personal risk factors . < 4 years . Lymphoma . D.M. . Renal failure . Malnutrition - Environmental risk factors (environmental exposure to TB)	No personal or environmental risk factors

- Treatment:

Short course of treatment gives better compliance and drugs combination decrease the risk of resistance, so the following protocol is recommended:

A- <u>Positive PPD in otherwise normal child (clinically and after investigation)</u>

Start INH alone for 3 months (as prophylaxis) then re-evaluate the patient:

1. No disease continue INH for 6 months
2. Sign of disease: treat as in B

B- <u>Diseased patient</u>: Full treatment:

1. INH: 6 months
2. Rifampicin: 6 months
3. Pyrazinamide: 2 months
4. In severe infections: (meningitis, milliary) give in addition Streptomycin (in patients < 8 years of age) or Ethambutol in patients > 8 years of age for 2-3 weeks

N.B. In Renal, Bone and CNS infection, treatment should continue for up to12-18 months.
Before starting therapy and during follow up CBCs, liver functions tests should be monitored.
Tuberculosis is a reportable disease, report to the health authority.

MALARIA

- Clinical manifestation:

History of travel to endemic area
Fever, chills, rigors, sweat, headache, pallor, jaundice, hepatosplenomegaly.

- Diagnostic test:

Stained blood film:
Thick blood film: for parasite identification
Thin blood film: for species identification
Parasitic index: % of infected RBCs
< 1%: mild parasitaemia
1-5%: moderate parasitaemia
> 5%: severe parasitaemia

- Treatment:

1. Supportive therapy: for fever, fluid and electrolytes
2. Specific therapy

A. Oral therapy: all types of malaria provided that P. Falciparum is susceptible.

- Chloroquine phosphate: 10 mg/kg stat, then 5 mg/kg 6 hours later, then 5 mg/kg/day for 2 consecutive days.

- If no response (no decrease in the parasitaemic index in 24 hours) after consultation of infectious disease department give Fansidar (Pyrimethamine and sulphadoxine) as single dose. If no response → Mefloquine hydrochloride as single dose (not in children < 15 kg weight).

B. Parenteral therapy: For those who have persistent vomiting or who are in coma.

- Quinine dihydrochloride, if not available
- Quinidine gluconate, if not available
- Chloroquine hydrochloride
- Parenteral therapy should be replaced by oral therapy as soon as possible.

C. Prevention of relapses (plasmodium vivax or p. ovale): → Primaquine phosphate for 14 days starting in the 3rd day of chloroquine phosphate.

- Chemoprophylaxis:

Starting day: 1 week before traveling to endemic area
End day: 8 weeks after leaving the endemic area
Drugs: Chloroquine is generally preferred: 5 mg/kg/once per week
For chloroquine resistant area other drugs can be used like: proguanil or fansidar.

Pertussis (Whooping cough)

Etiology: Bordetella pertussis (Gram-negative bacilli).
Incubation period: 6 - 20 days
Clinical features:

- Catarrhal phase (1-2 wk): rhinorrhea.
- Paroxysmal phase (2-4 weeks or longer): Bouts of coughing in runs of 10 or more followed by whoop and ends by vomiting.
- Convalescent phase (1-2 weeks): coughing slowly subsides over period which can last up to 3 months (hundred days cough as used to be said).

Complications:

- Respiratory: Pneumonia (more commonly by secondary bacterial infection), apnea, atelectasis, otitis media, sinusitis, pneumomediastinum, pneumothorax, interstitial or subcutaneous emphysema. Later on: Bronchiectasis.
- Hemorrhage: Epistaxis, retinal, subconjunctival, intraventricular.
- Hernia: Inguinal, umbilical, rectal prolapse, rupture of diaphragm.
- Cerebral anoxia: following apnea manifests as fits - 2.5%, encephalopathy 0.5%, apnea and sudden death may occur during very severe paroxysm.

Treatment:

- Erythromycin, when given within 14 days of onset of the disease, may eliminate the organism from nasopharynx, improve symptoms and reduce communicability.
- In severe paroxysmal attacks sulbutamol nebulization and steroid might be helpful.
- Treat complications.

Isolation & infectivity:

- The patient should be placed in droplets precautions for at least 5 days
- Treat with erythromycin for a total of 14 days.
- Close contacts of less than 7 year of age who are unimmunized or who have received fewer than 4 doses of DTP should have pertussis vaccine according to the recommended DTP schedule and erythromycin for 14 days.

COMMON CHILDHOOD INFECTIOUS DISEASES

Disease	Incubation period	Clinical picture	Complications	Isolation & Infectivity
Measles **Etiology:** RNA virus, (Paramyxo-virus)	8-12 days from exposure to the onset of symptom	Fever, malaise, coryza, cough, conjunctivitis, Koplick's spots. Skin rash (macular - maculopapular), starts from hairline then to the face, spreading down to involve the whole body and disappear in the same sequence.	Respiratory complications: Otitis media, pneumonia, laryngitis, tracheitis, bronchitis. Neurological complications : Encephalitis, in 0.1% within one week of the appearance of rash. SSPE (subacute sclerosing panencephalitis) 1:100,000 cases, occurs years after measles. Fatal in 6-12 months, Guillain-Barre syndrome, Hemiplegia. Other Complications: Hemorrhagic or "Black" measles (severe form), thrombocytopenia, appendicitis, keratitis myocarditis, optic neuritis, and progression of tuberculosis. **Treatment:** Symptomatic. Vaccine can prevent or ameliorate the disease if given shortly in the incubation period.	From 2 days before up to 5 days after appearance of the rash.

Disease	Incubation period	Clinical features	Complications	Isolation & infectivity
Rubella **Etiology:** RNA virus (Togaviridae)	14 -21 days	Asymptomatic infection is common. Mild coryza, erythematous maculopapular discrete skin rashes usually fade after 3 days. Lymphadenopathy: (most common), suboccipital, post-auricular, and cervical, are the most prominent. Splenomegaly ±	Not common in acquired rubella. Arthralgia, arthritis, Encephalitis (rare), Thrombocytopenia **Treatment:** symptomatic	Isolation is required for 7 days from the onset of the rash. Infants with congenital rubella should be considered contagious until one year of age unless nasopharyngeal and urine culture after 3 months of age are repeatedly negative.

Disease	Incubation period	Clinical features	Complications	Isolation & infectivity
Mumps **Etiology:** (RNA Paramyxo virus)	14 - 21 days	May be asymptomatic. Malaise, fever, headache, anorexia Later: parotid gland enlargement, unilateral or bilateral.	Glandular: Orchitis in post pubertal males (sterility is rare), epididymitis, oophoritis, pancreatitis, thyroiditis, mastitis. Non glandular: Aseptic meningitis, Encephalitis, Nerve deafness (usually unilateral), Nephritis, Myocarditis, Arthritis, Thrombocytopenia. **Treatment**: Symptomatic	9 days after the onset of parotid swelling

Disease	Incubation period	Clinical features	Complications	Isolation & infectivity
Infectious Mononucleosis **Etiology:** Ebstein-Barr virus (EBV)	30 - 50 days	Age-related, and range from asymptomatic to fatal infections. Prodrome: malaise, fatigue, headache, abdominal pain (lasting 1-2 wk). Typical symptoms and signs: Pharyngitis (with enlarged tonsils and exudates). Maculopapular rash (can be induced by ampicillin). Fever, Lymphadenopathy, Splenomegaly, Hepatomegaly, Palatal petechiae.	Gastrointestinal: Hepatitis: (considered part of the disease), Splenic rupture (avoid contact sports). Hematological: Hemolytic anemia (coombs +ve), thrombocytopenia, neutropenia, hemophagocytic syndrome, aplastic anemia. Neurologic: Seizures, ataxia, aseptic meningitis, Bell's palsy, encephalitis, Guillain-Barre syndrome. Respiratory: Upper airway obstruction (This is an emergency situation; give steroid and be ready for intubation), Interstitial pneumonia. Cardiac: Myocarditis. Malignancy: Burkitt lymphoma (African), Nasopharyngeal carcinoma (Chinese). **Treatment**: Symptomatic.	There are no measures to prevent EBV infection. Patients with recent EBV infection should not donate blood.

Disease	Incubation period	Clinical features	Complications	Isolation & infectivity
Chicken Pox **Etiology:** (varicella zoster) DNA Herpes-virus.	10 - 21 days, (usually 15 days)	Begins with fever and malaise. Rash starts as pruritic small macules, evolve within few hours to papules and then vesicles. Vesicles ulcerate, crust and heal, new crops appear for 3-4 days, usually beginning on the trunk and then the head, face and less commonly extremities. Severe neonatal varicella can occur when mother had the disease 5days before till 2 days after delivery	Secondary bacterial infections (staphylococcus, group A streptococcus). Isolated cerebellar ataxia (cerebellitis), has excellent prognosis without treatment. Encephalitis 1:1000, prognosis is guarded, may respond to acyclovir. Reye syndrome (avoid aspirin). Thrombocytopenia: Hemorrhagic chicken pox or bleeding Pneumonia (healed as microcalcification) Disseminated varicella can be fatal in immunocompromised children e.g. (leukemia, nephrotic syndrome). **Treatment**: Symptomatic Acyclovir for immunocompromised patients, Disseminated VZ. Vaccination for varicella is available now from the age of 1 year.	Till all skin lesions crusted (6th day after onset of rash).

VIRAL HEPATITIS

FEATURES	HEPATITIS A	HEPATITIS B	HEPATITIS C	HEPATITIS D	HEPATITIS E
Virus	HAV	HBV	HCV	HDV	HEV
Genome	RNA	DNA	RNA	RNA	RNA
Incubation Period	15 - 50 days	45 - 160 days	2-24 weeks	2 - 8 weeks	15 - 60 days
Onset	Usually acute	Usually insidious	Usually insidious	Usually acute	Usually acute
Transmission: - Oral (fecal) - Parenteral - Other	Usual Rare Food or water-borne	No Usual Intimate contact Perinatal	No Usual Intimate contact and perinatal: rare	No Usual Intimate contact less common	Usual No Water - borne
Sequelae: - Fulminant liver failure - Carrier status - Chronic hepatitis	Rare No No cases reported	Uncommon unless coinfected with delta. Yes Yes	Uncommon Yes Yes	Yes Yes Yes	Yes (in pregnancy) No No cases reported
Mortality	0.1%-0.2%	0.5% - 2%	1% -2%	2% - 20%	1% - 2% 20% in pregnant women

PAEDIATRIC EMERGENCIES & INTENSIVE CARE

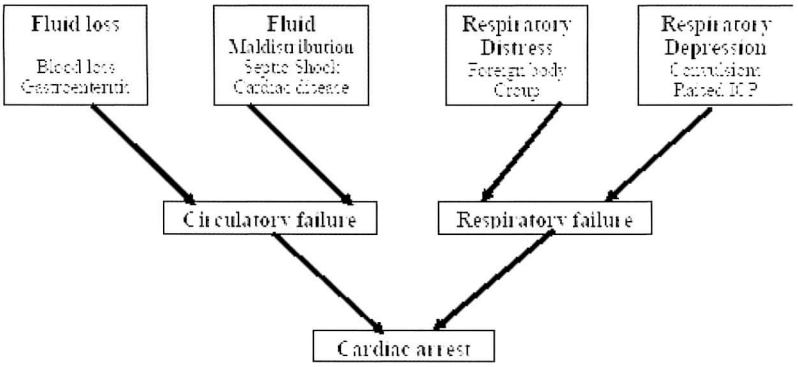
Critical illness in a child is a clinical state that may result in respiratory or cardiac arrest or severe neurological complication that required rapid cardiopulmonary assessment. In these cases, a quick **assessment** is foremost important to find out the need for immediate resuscitation.

Normal vital signs in infants and children [4,5]

Age	Heart rate	Blood pressure	Respiratory rate
1 month	100-180	85/50	30-60
6 months	120-160	90/53	30-60
1 year	90-140	91/54	20-40
2 years	80-140	91/55	20-30
6 years	75-100	96/57	20-25
10 years	60-90	102/62	17-22
12 years	55-90	107/64	17-22

Cardiac arrest in infants and children is rarely a sudden event. It is usually preceded by progressive deterioration in respiratory and circulatory function. So impending cardiac arrest may be averted by early recognition of respiratory failure, circulatory failure and neurological failure.

Infants and young children have less respiratory reserve than older children or young adults, and many patients admitted to the PICU have either a primary respiratory disorder or a respiratory component to their disease. Respiratory failure is the commonest cause of cardiac arrest in children. It may result from upper or lower airway disease. Tachypnoea, recession, use of accessory muscles, grunting are all signs of increased work of breathing. Inspiratory stridor is a sign of upper airway obstruction while wheezing denotes lower *airway* disease. The onset of fatigue, or coincident neurological impairment, may diminish these important signs and produce a false impression of well being



Assessment of adequacy of breathing and Circulation

Work of breathing
- Recession
- Respiratory rate
- Inspiratory or Expiratory noises
- Grunting
- Accessory muscle use
- Flaring of the nostrils

Effectiveness of breathing
- Breath sounds
- Chest expansion
- Abdominal excursion

Cardiovascular status
- Heart Rate
- Pulse volume
- Capillary refill
- Blood pressure

Effects on other organs
- Respiratory rate and character
- Skin appearance and temperature
- Mental status
- Urinary output

Assessment of Conscious level

A Alert
V Response to voice
P Response to pain
U Unresponsive

Cardio Pulmonary Resususcitation in children

A respiratory or cardiac arrest in infants and children is an acute, life-threatening event requiring immediate intervention. Cardiopulmonary resuscitatation is the restoration of automatic and effective breathing and circulation. There are two stages of intervention in CPR:

1. **Basic Life Support:** The restoration of effective ventilation and circulation using non-invasive methods. That is, breathing expired air into the lungs without mechanical devices and using closed cardiac compression techniques.

A: For Assessment:

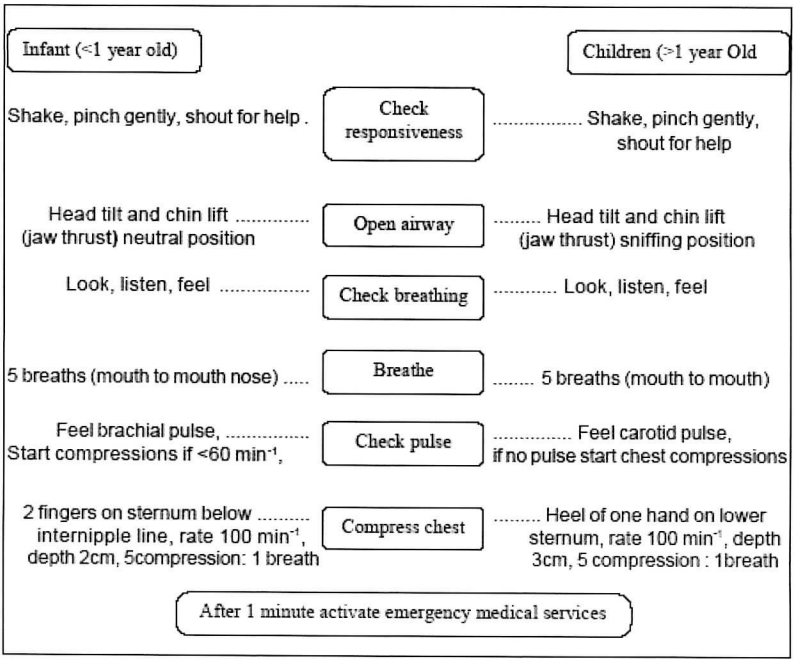
- Assess responsiveness – shake (avoid shaking neck if trauma suspected), pinch gently if no response & shout for help.

B: For breathing:

- Open airway: Head tilt and chin lift or jaw- thrust (in case of suspected neck trauma)
- Check breathing: Look to respiratory movement, listen breathing sound, and feel flow of air by your cheek, if no breathing.
  - Start breathing –
    - 1st 2 breaths: observe for visible chest expansion
      - < 1 year give mouth to mouth and nose breathing
      - 1 year give mouth to mouth breathing closing the nostril by
    - Fingers

C: For Circulation –

- Check pulse: Brachial < 1 year
  Carotid > 1 year
- Start chest compression at lower half sternum or one finger below nipple line in infants.
  < 1 year give 100 compressions/min: depth 2 cm
  > 1 year give 100 compressions/min: depth 3 cm
  Old child: depth 4-5 cm or $^1/_3$ - ½ depth of chest.
  Give 1 breath/ 5 compressions in general
  For neonate give 120 compressions/min & 1 breathe for every 3 compressions. Old child (>8yrs) may also be given 2 breaths/15 compressions as adult.
  For compression use 2 fingers (index and middle finger) or both thumbs encircling the chest for neonates & small babies, and use heel of one hand for younger children, and heel of 2 hands for older children.
  Continue BLS until advanced life support can be offered or if no response after 30 minutes. Activate advanced life support team after 1 minute & continue resuscitation.



2.ADVANCED LIFE SUPPORT(ALS):Remember by letters **A & B, C, D, E, F.** : Invasive procedures aimed at restoration of ventilation and circulation. These include bag-valve-mask ventilation, endotracheal intubation, intravenous, introsseous and /or endotracheal drug administration

A & B: Airway & Breathing:

- Intubate and ventilate with 100% O_2, alternatively use Ambu bag and mask for ventilation.

C: Circulation:

- Continue cardiac massage same as in BLS.

D: Drugs:

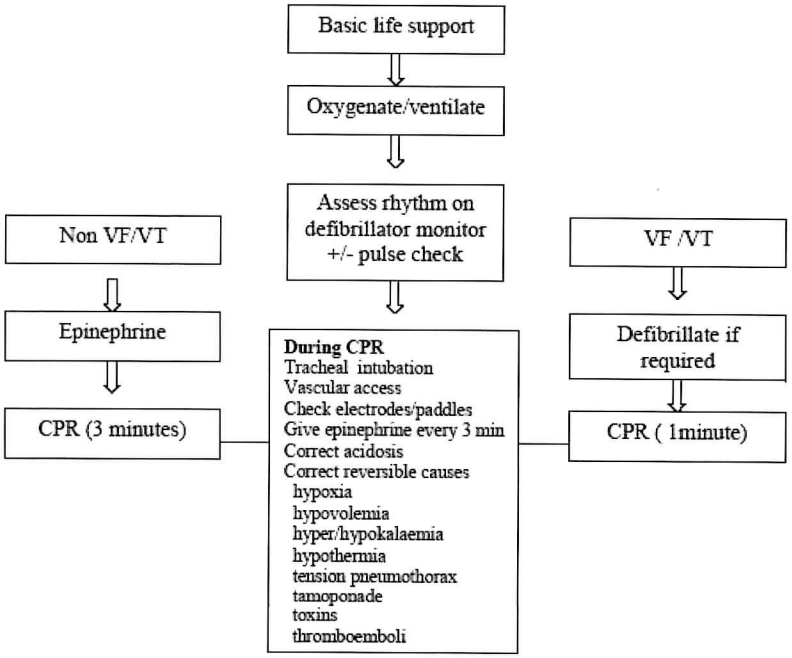
- Intravenous or Intraosseous line for drugs
  Drugs used most commonly for ALS.
  a) Adrenaline: (0.1 ml/kg of 1 in 10,000) IV or via ET tube (10 times of IV dose via ET tube). Dose of adrenaline may be increased if there are recurrent subsequent arrests.
  b) Bicarbonate 1 mmol/kg of 8.4% IV (dilute in 5% dextrose 1:1), Should not be given until ventilation is established.
  c) Atropine 0.02 mg/kg (maximum 0.6 mg) IV or via ET tube (used mainly for vagal induced bradycardia)
  d) Other ionotropic agents, antiarrhythmic drugs, DC shock, colloids and crystalloids are used in special circumstances subsequently.

E: Evaluation:

- Evaluate the success of CPR by clinical examination: When there is spontaneous cardiac output feel for pulse if pulse& BP good, stop cardiac massage & monitor ECG. Evaluate the success of CPR further by some essential laboratory investigations.
- **Investigations:**
    - Arterial blood gas, complete blood count, urea, creatinine, glucose,
    - Electrolytes, liver functions, chest x-ray, ECG.

F: Further steps & follow up:

Inform family about cardiac arrest & resuscitation.
Proceed for further history, examination and management. Decide about fluid therapy. Look for urine output. Consider ionotropic support by dopamine and/ or dobutamine 2-20 µgm/ kg/min. Consider need for further ventilatory management. Maintain body temperature. Find and treat underlying cause of cardiac arrest. Follow up for neurological impairment.



RESPIRATORY FAILURE

Definition:
Respiratory failure is said to occur when the respiratory system is unable to deliver oxygen to and/or remove carbon dioxide from pulmonary circulation, there by leading to hypoxemia and/or hypercapnia.

Classifications:
a) Acute & chronic
b) Type 1 & type II
Type I: Low arterial PaO_2 (hypoxemia) & normal or low $PaCO_2$
Type II: Low arterial PaO_2 (hypoxemia) & elevated $PaCO_2$ (hypercapnia)

Clinical Features:
Features due to hypoxemia:
Cyanosis, pallor, restlessness, irritability, tachycardia, increased respiratory efforts evident by: active alae-nasi; intercostals; subcostal; suprasternal retractions; grunting, etc.
Features due to hypercapnia:
Sweating, tremor, warm extremities, bounding pulses, hypertension, bradycardia, headache, mental confusion, seizure, coma etc.
Other features: may be related to the underlying cause of respiratory failure.
Diagnosis of respiratory failure is to be confirmed by arterial blood gas estimation.
Etiology of Respiratory Failure: Respiratory failure may result from-

a) Obstructive respiratory diseases:
   - Neonates and young infants: Choanal atresia, meconium aspiration, bronchiolitis etc.
   - Older infants & children: bronchial asthma, adenotonsillar hypertrophy Bronchopneumonia, foreign body inhalation, epiglottitis etc.

b) Restrictive respiratory diseases:
   - Neonates & young infants: hyaline membrane disease, Pulmonary hypoplasia, diaphragmatic hernia, congenital lobar emphysema etc.
   - Older infants & children:
     - Pulmonary: pneumonia, pneumothorax, plural effusion etc.
     - Neurological: Depression of respiratory centre from poisoning, injury or raised intracranial pressure, or weakness of respiratory muscles (poliomyelitis, Guillain-Barre syndrome, myasthenia gravis, myopathies etc.).
     - Mechanical: severe obesity, kyphoscoliosis, chest wall injury etc.

c) Inefficient gas transfer:
   - Pulmonary edema, carbon monoxide poisoning, embolism, severe anemia, Right to left shunt..

Approach to Management:
Severest from of respiratory failure may present with cardiopulmonary arrest.
Management in such cases should be according to the guidelines of cardiopulmonary resuscitation. Most of the respiratory illnesses which presents with respiratory failure come to the hospital before arrested. Following is a general guideline for their management.

1. **Oxygen therapy:**

Oxygen may be given by; nasal cannulas, nasal prongs, face mask, or by head box. Many patient's with Type I respiratory failure will improve with oxygen therapy while proper treatment for the underlying cause is provided. Oxygen therapy may be guided by pulse oxymetric monitoring. Give enough oxygen to keep SpO_2 (O_2 saturation) around 95%. SpO_2 of less than 90% while patient on high inspired oxygen may indicate the need for more aggressive steps.

2. Further respiratory support:

If despite of oxygen therapy and adequate treatment of underlying cause the patient continues to deteriorate, following respiratory supports should be considered:

I. Continuous positive airway pressure (CPAP)
II. Artificiel ventilatory support (traditional artificiel ventilation)
III. High frequency ventilation
IV. Extra corporeal membrane oxygenation (ECMO)

3. Investigational therapy: Liquid ventilation, Nitric oxide inhalation.

Artificial ventilatory support:

Present day most of the artificial ventilators deliver air and oxygen to the patients by positive pressure. Institution of intermittent positive pressure ventilation (IPPV) requires endotracheal intubation. Size of the endotracheal (ET) tube varies with age of the child.

At birth:	<1 kg. birth weight	2.5 mm.
	> 1 kg. birth weight	3.0 mm.
1 - 6 months		3.5 mm.
6 - 12 months		4.0 mm.
12 - 18 months		4.5 mm

After 2 years use the following formula or select a size comparing the little finger of the patient.

$$\text{ETT Size} = \frac{\text{Age in years}}{4} + 4\text{mm (3mm for cuffed tube)} \pm 0.5\text{mm.}$$

Changes of ventilator settings Suggested in Hypoxemia and *Hypercapnea*:

If the patient is hypoxemic:

1. Increase FiO_2 (Fractional Inspiratory Oxygen or O_2%
2. Increase PEEP (Positive End Expiratory Pressure)
3. Increase PIP (Peak Inspiratory Pressure), MAP(Mean Airway Pressure), I:E (Inspiratory Expiratory ratio)
4. Increase Rate of Respiration or Frequency

If patient is hypercapnic:

1. Increase Rate
2. Increase PIP
3. Reduce PEEP and I:E

Every change in ventilator settings is to be followed by blood gases to see the desired effect. One or two changes should be made at a time. Consider the possible complications of changes of ventilator setting to the patient.

Complications of artificial ventilation:

A) Respiratory:
- Complications related to intubation: Cardiac arrest during the procedure from vagal stimulation, injury to nose and mouth, sinusitis, otitis media, subglottic stenosis.
- Barotrauma - pneumothorax, pulmonary interstitial emphysema, pneumomediastinum
- Complications of oxygen therapy: Retinopathy of prematurity, adult respiratory distress syndrome, bronchopulmonary dysplasia etc.

B) Circulatory:
- Reduced venous return, reduced cardiac output, systemic hypotension.
- Impaired venous return from high PEEP and high mean airway pressure may increase venous congestion in the brain causing raise of intracranial pressure and chance of intracranial hemorrhage.

C) Complications from medications which are used for assisted ventilation

Checklists for a patient who suddenly deteriorates on artificial ventilation:

Remember by the abbreviation **DOPED:**

1. The tube may be **displaced** from trachea to the pharynx and esophagus. During bagging air will go to the stomach which may be heard over epigastrium by the stethoscope, abdomen will be distended. Remove and replace the tube to the trachea.
2. The tube may be **obstructed** by thick mucous secretions. You will feel high resistance during Ambu bagging. Chest wall will not move. Absent breathe sounds on both sides. The endotracheal tube is to be removed and replaced by another new tube. You can see the mucus plug in the previous ET tube.
3. Patient may have developed **pneumothorax**. Breath sounds will be absent or reduced on the side of pneumothorax. Percussion note will be hyper-resonant. Bed side transillumination will be positive in neonates. Patient needs intercostal drainage.
4. **Equipment failure:** Check the ventilatory circuit, a tube might have been accidentally disconnected. Disconnect the patient from ventilator and ventilate by Ambu-bag and oxygen, patient will improve. Connect again the disconnected tube
5. The pathological process might have been seriously **deteriorated**.

Acute Respiratory Distress Syndrome

Definition:
Acute respiratory distress characterized by acute lung injury, noncardiogenic pulmonary edema and severe hypoxia.

Diagnostic Criteria

1. Identifiable associated condition
2. Acute onset
3. Pulmonary artery wedge pressure < or = to 18mm or absence of evidence of left atrial hypertension
4. Bilateral infiltrates on chest radiography
5. Pao2/Fio2 ratio < or = to 200
   *[Pao2/Fio2 ratio < or = to 300 is defined as Acute Lung Injury]
   -American-European Consensus Connference Statement,1994

Risk Factors:

Pulmonary	**Extra-pulmonary**
Bacterial pneumonia	Sepsis
Viral pneumonia	Trauma
Aspiration	Multiple transfusion
Near Drowning	Peritonitis
Inhalation injury	Cardiopulmonary bypass

Pathophysiology:

1. Direct lung injury or systemic insult occurs
2. Release of pro-inflammatory agents i.e. TNFα, interleukins
3. Migration of neutrophils producing oxygen radicals and proteases
4. Endothelial and epithelial cell damage leads to increased permeability and the influx of fluid into the alveolar space. (pulmonary ARDS—epithelial damage, extra-pulmonary ARDS—endothelial damage initially)
5. Surfactant is abnormal
6. Impaired fibrinolysis leads to capillary thrombosis/microinfarction

Pathology of ARDS

Exudative Phase (Day 1-7)	Proliferative Phase (Day 7-21)	Fibrotic Phase (> Day 21)
Interstitial and intra-alveolar edema	Interstitial myofibroblast reaction	Collagenous fibrosis
Hemorrhage	Lumenal organizing fibrosis	Microcystic honeycombing
Leukoagglutination	Chronic inflammation	Traction bronchiectasis
Necrosis-Type I Pneumocytes	Parenchymal necrosis	Arterial tortuosity
Hyaline membranes	Type II pneumocyte hyperplasia	Mural fibrosis
Platelet-fibrin thrombi	Obliterative endarteritis	Medial hypertrophy
	Macrothrombi	

Tomashefski, J. *Acute Respiratory Distress Syndrome*. Clinics in Chest Medicine: 21(3) Sept. 2000.

Evaluation:
physical exam: tachypnea, tachycardia, altered mental status
blood gas montoring: initial **respiratory alkalosis** may precede infiltrates, later: alveolar edema → VQ mismatch/shunt → **severe hypoxia**

Imaging: CXR progression from diffuse interstitial infiltrates to diffuse, fluffy alveolar spaces; later reticular pattern suggests interstitial fibrosis. CT demonstrates dependent (posterior if supine) infiltrates and atelectasis, with anterior hyperinflation.
*most patients with ARDS develop diffuse alveolar infiltrates and progress to respiratory failure within 48 hours of the onset of symptoms

Treatment:
1. Treatment of underlying cause or associated condition
2. Ventilatory support- ensure "adequate" oxygenation/ventilation while minimizing ventilator induced lung injury.
   Avoid over or under-inflation: Usually this requires PEEP of 6-12, depending on severity. Remember things tend to get worse before they get better-it is not unusual for patients to require increasing PEEP as their disease worsens.
   Use low tidal volumes, 5-7 cc/kg. Monitor peak pressure (PIP or plateau pressure). It should be less than 30-35.
   Use longer inspiratory times than usual for age.
   Tolerate hypercapnia, monitor pH, try to keep >7.2
   Tolerate hypoxemia if necessary to keep FiO2 <60%. If on <60%, Sat goal should be ~92, if not able to maintain 92 on <60%, tolerate 88%.

Monitor trends closely—absolute numbers are not usually important, trends in numbers are often extremely important.
Remember that cardio-pulmonary interactions occur, and ventilator maneuvers may effect hemodynamics.

3. Pharmacologic treatment- no proven role as yet. Drugs sometimes used include steroids (late phase), NitricOxide (no proven survival benefit),
4. Consider high-frequency oscillatory ventilator
5. Consider prone position
7. Monitor and/or Prophylaxis for complications-
    a. GI bleed.
    b. Thromboembolism
    c. Nosocomial infection
6. Supportive care- nutrition, sedation/pain control
7. Consider ECMO if no improvement

Complications:

Barotrauma- pneumothorax, pneumomediastium, subcutaneous emphysema
Cardiac- hypotension
GI- stress-related gastrointestinal hemorrhage
Death estimated to occur in 20 (low risk)-90% (highest risk, BMT) of cases. Previously well children and those with extrapulmonary ARDS have a better prognosis. The mortality from ARDS has fallen significantly since it was first described in 1967.

Diabetic Ketoacidosis

Definition:

metabolic acidosis
ketonuria/ketonemia
hyperglycemia (not mandatory)
dehydration
associated electrolyte disturbances: psuedohyponatremia, hypokalemia, hypophosphatemia
PICU admission criteria: (depends on the case/Hospital policy)
PH<7.25, HCO3 <15, mental status changes, cardiac arrhythmia
Insulin infusion that requires titration

Pathophysiology:

1. Occurs due to an absolute or relative insulin deficiency along with an excess of counterregulatory hormones (e.g. glucagon, catecholamines, cortisol, and growth hormone) as seen with infection or stress results in stimulation of lipolysis and increased levels of circulatory free fatty acids
2. Fatty acids are oxidized in liver resulting in elevated levels of circulating ketone bodies (beta-hydroxybutyrate and acetoacetate)

3. Counterregulatory hormones stimulate hepatic ketogenesis as well as gluconeogenesis and glycogenolysis resulting in excess glucose production and hyperglycemia
4. DKA can occasionally present without hyperglycemia such as during pregnancy, in patients who have been partially treated and those with prolonged vomiting with little to no carbohydrate intakeas blood glucose rises, the ability of the proximal tubule to resorb glucose is exceeded and glycosuria occurs resulting in osmotic diuresis and dehydration

Evaluation:

1. Careful history: vomiting, abdominal pain, polyuria, polydipsia, nocturia, weakness, heavy breathing or shortness of breath, symptoms of intercurrent illness, mental status changes, sweet odor to breath, weight loss.
2. Physical exam: dehydration (dry mucous membranes, poor skin turgor, poor pefusion), tachycardia, hypotension, Kussmaul respirations, somnolence, hypothermia, impaired consciousness
3. Lboratory studies:
   venous blood gas
   metabolic panel/blood glucose
   urine or serum ketones
   complete blood count
   anion gap
   consider: HgA1C, TSH, freeT4
   other signs of infection i.e. urinalysis/culture

Useful Equations:

Correction for psuedo/dilutional hyponatremia:
Na^+ (corrected) = Na^+ (measured) + [(serum glucose – 100)/100] x 1.6
Anion gap: [(Na^+ + K^+) – (HCO_3^- + Cl^-)]

Treatment:

1. ABC's → ensure adequate airway, ventilation and circulation
2. Correct fluid deficits
   calculate fluid deficit (may assume 5-10% dehydration)
   i.e. total fluid deficit = 10ml/kg for each 1% dehydrated
   replace evenly over 48 hours in addition to maintenance fluids
3. Correct electrolyte deficiencies
   consider normal saline or 1/2 normal saline
   potassium shifts extracellularly due to acidosis- therefore despite normal serum potassium levels a total body deficit usually exists if serum K < 5, replace with 40 mEq potassium in fluids initially. You may need to add more replace hypophosphatemia by using Kphos for 1/2of potassium replacement
   example fluids: NS + 20 mEq KCl/L + 20 mEq Kphos/L
4. Correct metabolic acidosis by interrupting ketone production
   begin with continuous insulin drip 0.05- 0.1 units/kg/hr IV
   start with lower dose and titrate to achieve glucose drop no more than 50-100 mg/dL/hour.

monitor blood glucose q1-2 hours → when glocuse reaches 250-300 mg/dL add D5 to fluids, change to D10 (try to increase dextrose in IVF's to keep blood sugar 200-300 rather than decreasing rate of insulin drip until acidosis is corrected)monitor venous blood gas and electrolytes q2-4 hours until out of DKA
monitor urine for ketones and glucose with each void
when acidosis resolved (HCO3 >18), pt tolerating PO and mental status normal consider switching to SQ insulin = 0.5-1.0 unit/kg/day
2/3 total dose in am (1/3 Regular, 2/3 NPH)
1/3 total dose in pm (1/2 Regular at dinner, 1/2 NPH at bedtime)
(please consult the endocrinologist if available)

5. Assess for and treat any underlying causes for DKA (e.g. infection)
6. Cclosely monitor for and treat any complications of DKA

Complications:

1. Cerebral edema- the leading cause of mortality; occurs in 1-2% of children with DKA; risk factors include rapid shifts in osmolality, excessive fluid administration, use of hypotonic fluid; symptoms include declining/fluctuating mental status, symptoms of increased intracranial pressure such as dilated or unequal pupils, Cushings triad. *Treatment*: Mannitol, consider intubation, mechanical ventilation
2. Cardiac arrythmia- due to electrolyte disturbance (hypo/hyperkalemia)
3. Fluid overload
4. Hypoglycemia

Status Epilepticus

Definition:

1. A life-threatening medical emergency defined as frequent or prolonged epileptic seizures
2. Many definitions including a continuous seizure lasting longer than 30 minutes or repeating convulsions lasting 30 minutes or longer without recovery of consciousness between them. Current thinking involves shorter periods of time.
3. onset may be partial or generalized

Epidemiology:

1. A common neurologic medical emergency, affecting 65,000 to 150,000 persons in the United States yearly
2. estimated that 1.3-16% of all patients with epilepsy will develop SE at some point in their lives (in some may be the presenting seizure)
3. more common in childhood than in adults, no sexual predominance
4. mortality rate is as high as 10%, rising to 50% in elderly patients
5. many possible etiologies as listed below:

Causes of Status Epilepticus

Background of Epilepsy

- Poor compliance with medication
- Recent change in treatment
- Barbiturate or benzodiazepine withdrawal
- Pseudostatus epilepticus
- Underlying infection/fever

Presenting de novo

1. Recent stroke
2. Meningo-encephalitis, meningitis, encephalitis
3. Acute head injury
4. Cerebral neoplasm
5. Demyelinating disorder
6. Metabolic disorders (e.g. renal failure, hypoglycemia, hypercalcemia)
7. Drug overdose (e.g. TCA's, phenothiazines, theophylline, isoniazid, cocaine)
8. Inflammatory states (e.g. systemic lupus erythematosus)
9. New onset seizure disorder

Evaluation and Treatment:

1. Eevaluate and support ABC's
2. Obtain IV access if possible
   - Check glucose
   - If access, draw: lytes including Ca/Mg, Bun/Cr, LFTs
   - Consider CBC/blood cx if infection possible
   - Draw anticonvulsant levels, tox screen if indicated
3. Administer a rapidly acting benzodiazepine
   - If IV: lorazepam (ativan) 0.5mg-1mg/kg (max 10mg)
   - May repeat ativan
   - Administer long acting AED

   Fosphenytoin 15-20 mg/kg IV or
   Phenobarbitol 15-20 mg/kg IV
4. If seizures persist, consider additional ativan or additional bolus of phenytoin or phenobarb 5mg/kg IV
5. ABC's…continue to eval; may need intubation if not able to manage airway
6. Last resort may need to induce pentobarb or general anesthesia (propofol) coma after airway secured
7. Watch for potential complications including hypothermia, acidosis, hypotension, rhabdomyolysis, renal failure, infection and cerebral edema
8. Continue to search for and treat any underlying cause

Complications:

hypoxia
metabolic and respiratory acidosis

increased or decreased cerebral blood flow
hypo or hyperglycemia
rhabdomyolysis
hyperkalemia
hyperpyrexia
cardiac dysfunction, arrythmias, hypotension
permanent neurologic sequelae (e.g. motor deficits, MR, epilepsy)
death

Traumatic Brain Injury and Increased Intracranial Pressure

Definition:

increased intracranial pressure results when the volume of one of the cranial contents (brain parenchyma, cerebrospinal fluid, or blood) increases and adaptive measures are unable to compensate
increased ICP is dangerous when it compromises cerebral perfusion, leading to further cell damage, cerebral edema and eventual displacement and herniation of the brain
classification of brain edema:

vasogenic- characterized by increased permeability of brain capillary endothelial celsls, as in tumor, abscess, hemorrhage, infarction contusion, lead intoxication, and meningitis; the neurons and glia are relatively normal

cytotoxic- characterized by failure of the normal homeostatic mechanisms that maintain cell size: neurons, glia and endothelial cells swell; prominent in hypoxic-ischemic injury, osmolar injury, some toxins, and secondary injury following head trauma

interstitial- characterized by an increase in the water content of the periventricular white matter due to obstruction of CSF flow

Pathophysiology:

1. Te brain is composed of *three components*: brain (cells and intercellular fluid), blood and CSF; increases in the size of any of the three compartments can lead to increased ICP
2. PO2, pCO2, pH and blood pressure all effect cerebral blood flow, but may act differently in an injured brain compared to a normal brain
3. Increased pCO2 will cause an increase in cerebral blood flow, and hence an increase in ICP, low pO2 will also cause an increase in cerebral blood flow and ICP
4. Brain injury occurs in 2 phases: (1) the **primary injury** that occurs at the moment of impact and results from a transfer of kinetic energy to the brain and (2) the **secondary injury** that is a biochemical and cellular response to the initial trauma
5. The primary injury causes direct cellular damage; we cannot do anything to reverse the primary injury as neurons do not regenerate
6. The secondary injury is delayed, usu. peaking at 48-72 hours and occurs in response to the hypoxia, hypoperfusion and cell damage

that result from the initial trauma; our goal in management is to prevent, as much as possible, secondary injury

Causes of Brain Injury and Increased ICP

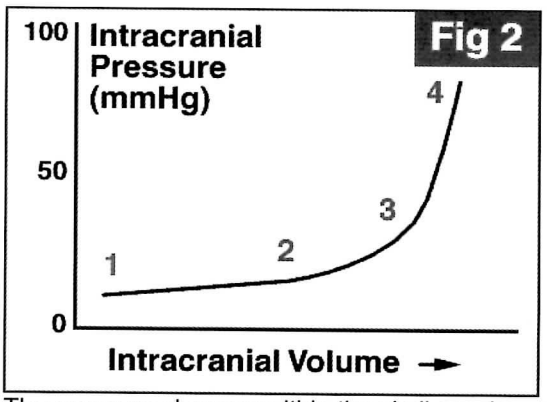
Generalized Brain Injury

- Hypoxic-ischemic injury
- Diffuse head injury i.e. shaken baby syndrome
- Osmolar injury (hypo-osmolality, hyerosmolality, DKA)
- Encephalopathies (Reye's syndrome, hepatic encephalopathy)
- Infection (meningitis, encephalitis)
- Toxins

Focal Intracranial Lesion

- Vascular: subdural, epidural, intraparenchymal hemorrhage, AVM
- Focal traumatic lesion, focal edema w/o bleeding
- Tumor
- Abscess

CSF Obstruction



The pressure changes within the skull are drawn in the classical curve which indicates an increase in volume with little change in pressure until a certain point is reached when a further small change in volume results in a large increase in pressure: 1-2 compensation phase; 3-4 decompensation phase

Evaluation:

1. Clinical history: -h/o trauma, symptoms including headache, vomiting, depressed level of consciousness i.e. confusion, restlessness, progressive unresponsiveness
2. Physical exam: abnormal posturing, abnormal breathing pattern, abnormal cranial nerve findings, papilledema, hypertension with bradycardia or tachycardia, bulging fontanelle
3. Cushing's triad: increased ICP, hypertension, bradycardia or tachycardia
   →bradycardia and Cushings's triad is a **late sign of increased**

4. Specific neurological Assessment to include Glasgow Coma Scale and:
    a. level of consciousness and mental status
    b. pupil size and shape and light response
    c. extraocular movements and visual acuity
    d. motor movement
    e. extremity strength
    f. headache, nausea, & vomiting
    g. fontanels, cranial sutures, & head circumference for pediatric patients < 2 years
    h. seizure activity

    Cerebral perfusion pressure (CPP) = MAP-ICP or MAP-CVP

Management:

1. Airway -remember avoid manipulation of neck in trauma; a child w/ a GCS <8 should be intubated as a general rule to protect the airway; use meds during intubation that will reduce the ICP response to intubation.
2. Breathing- ensure adequate oxygenation and avoid hypercapnia (mild hyperventilation is appropriate)
3. Circulation- provision of adequate cardiac output and blood pressure is essential; avoid lowering the osmolarity of blood (NO hypotonic fluids, normal saline or LR are good options as is albumin)
4. IV access and Lab evaluation: consider blood gas, electrolytes including Ca/Mg/Phos, Osmolality, blood glucose, LFT's ammonia, CBC/coags, toxicology screen, blood/urine/spinal tap
5. CT scan without contrast- evaluate for signs of trauma, bleed, edema
6. Evaluate and treat possible complications: hyperthermia, glucose abnormalities, seizures
7. Provide analgesia and sedation if indicated
8. Positioning- HOB elevated with head midline to avoid impeding venous return
9. Surgical management if indicated (drainage of blood, removal of tumor, drainage of CSF or shunt revision)
10. Intracranial pressure monitoring (intraventricular drain, intraparenchymal catheter (Camino), subarachnoid bolt). The goal is to maintain cerebral perfusion pressure 50-70 mmHg/ ICP <20, and detect "events": rebleed, herniation, etc.
11. Mechanical ventilation: sats >95%, avoid hypercapnia, consider short- term hyperventilation
12. Mannitol- decreases blood viscosity by lowering hematocrit, may reduce brain water content in the uninjured portion) → give rapidly, "chronic" dose is 0.25-0.5 mg/kg, in impending herniation give a large 1 gram/kg dose quickly; watch blood pressure and renal function
13. Lasix- synergistic in combo with mannnitol for reducing ICP

14. Other: barbiturates-controversial, steroids- will help reduce vasogenic edema (around tumors), no effect on cytotoxic brain edema or in the management of head trauma
15. Fluid Management- avoid hypotension and hypo-osmolality; look for SIADH; reasonable regimens include D51/2 NS or D5NS at slightly less than maintenance (so as to avoid Na overload) follow electrolytes and volume status closely. Do not restrict volume early in resuscitation.

Shock

Definition:

- Inadequate tissue perfusion to supply oxygen and nutrients to meet the metabolic demands of the body
- Three major types include *hypovolemic, distributive and cardiogenic*
- **Hypovolemic shock** is the most common form, and is due to an absolute loss of volume from the vasculature (blood loss (hemorrhage), body water loss (dehydration) or loss of plama)
- **Distributive shock** results when total ciruculating volume has been redistributed and a functional hypovolemic state results (seen in sepsis, neurogenic shock and anaphylaxis)
- **Cardiogenic shock** occurs when the heart is unable to maintain cardiac output (may be intrinsic i.e. heart failure or extrinsic i.e. tamponade)
- **Compensated shock** is the state of tissue hypoperfusion in which adaptive physiologic responses are still able to maintain blood pressure
- **Decompensated shock** is the state in which the adaptive physiologic responses can no longer compensated and central organ perfusion is no longer maintained as heralded by hypotension

Evaluation: rapid evaluation of airway, breathing and circulation

- **Clinical history**
  - underlying disease, recent infection or illness, trauma, surgery, etc.
- **Physical exam**
  - ABC's first, heart rate, blood pressure, respiratory rate, pulses, skin perfusion, altered mental status, decreased level of consciousness, urine output, other signs of trauma or focus for indication of infection
- **Studies**
  - Labs including CBC, CMP, blood gas, coagulation panel, blood culture; consider amylase in trauma, lactate
  - Imaging including chest xray, others in trauma (pelvis/abdomen)
  - Consider CT of head/abdomen if stable
  - Consider echocardiogram if possibility of cardiogenic shock
  - Urine and CSF studies in suspected septic shock

Treatment:
Aggressive supportive care is the primary goal in the ED
Airway

- Obtain and maintain
  - Beware of hemodynamic instability with intubation
  - Stress resuscitate, atropine
- Early intubation:
  - Neonates and young infants have *low FRC*

Breathing

- Oxygenation and adequate ventilation
- Pulmonary dysfunction occurs *early* and *often*
- Supplemental O2
- Continuous SaO2
- Serial ABG/VBG
  - Acidosis negative prognostic indicator

Circulation

- Intravascular access
- Resuscitation of circulation must be early and complete
  - *RAPID LARGE VOLUME BOLUSES OF IV FLUIDS*
  - 20 ml/kg iv/io q 5-10 minutes *(>/= 60ml/kg may be required)*
  - Titrated to:
    - *Measures of cardiac output*
      - *heart rate, urine output, capillary refill, level of consciousness*
    - *? Hepatomegaly*
- *Colloid or Crystalloid?*
  - *NS and RL are equally effective*
  - *colloids offer no additional benefit*
  - *No role for 2/3 1/3 or D5W*

  BP is not reliable indicator of the adequacy of resuscitation , the child's compensatory mechanism is to increase SVR and HR
- Markers of resuscitative effort:
  - Physiologic variables *e.g. tissue perfusion*
  - Serum lactate, HCO3-, CvO2/SvO2
- **Vasopressors/Inotropes**
- Only ***following*** aggressive volume resuscitation
- The septic child may present with:↓ CO, ↑ SVR ,or ↑ CO, ↓ SVR , or ↓ CO, ↓ SVR
- **Dopamine**
  - First line therapy
  - Immediate precursor of NE and E
  - β1 and dopaminergic agonist
  - AE = persistent tachycardia, and increased pulmonary pressures
- **Epinephrine**
  - Dopamine refractory
  - Infant (dopamine-insensitive)

- cold shock
- **SvCO2 < 70 %**
- Mixed α and β agonist
- AE = ↑oxygen consumption, ↑ lactate and ↓ splanchnic flow

- **Norepinephrine**
  - Dopamine refractory shock
  - warm shock
  - SvCO2 >/= 70 %
  - Mixed α and β agonist

- **Vasodilator therapy**

High SVR state despite aggressive volume resuscitation and inotropic support, cold shock, **SvCO2 < 70 %**

- Nitrosovasodilators
  - Epi resistant ↓ CO with ↑ SVR
- Inhaled NO
  - Neonate with PPHTN
- Phosphodiesterase inhibitors

THERAPEUTIC ENDPOINTS

PHYSICAL EXAM

- Normal mental status
- Capillary refill < 2 seconds
- Normal/symmetrical pulses
- U/O>/= 1 ml/kg/hr

LABORATORY

- Normalized lactate+/- HCO3-
- Cv02 > 70 %

TARGETED THERAPY

- Parenteral antibiotics
  - Early and appropriate
  - Cultures before when possible
- +/- Surgical drainage if indicated
- Acyclovir in septic neonate with suspect HSV

Steroids

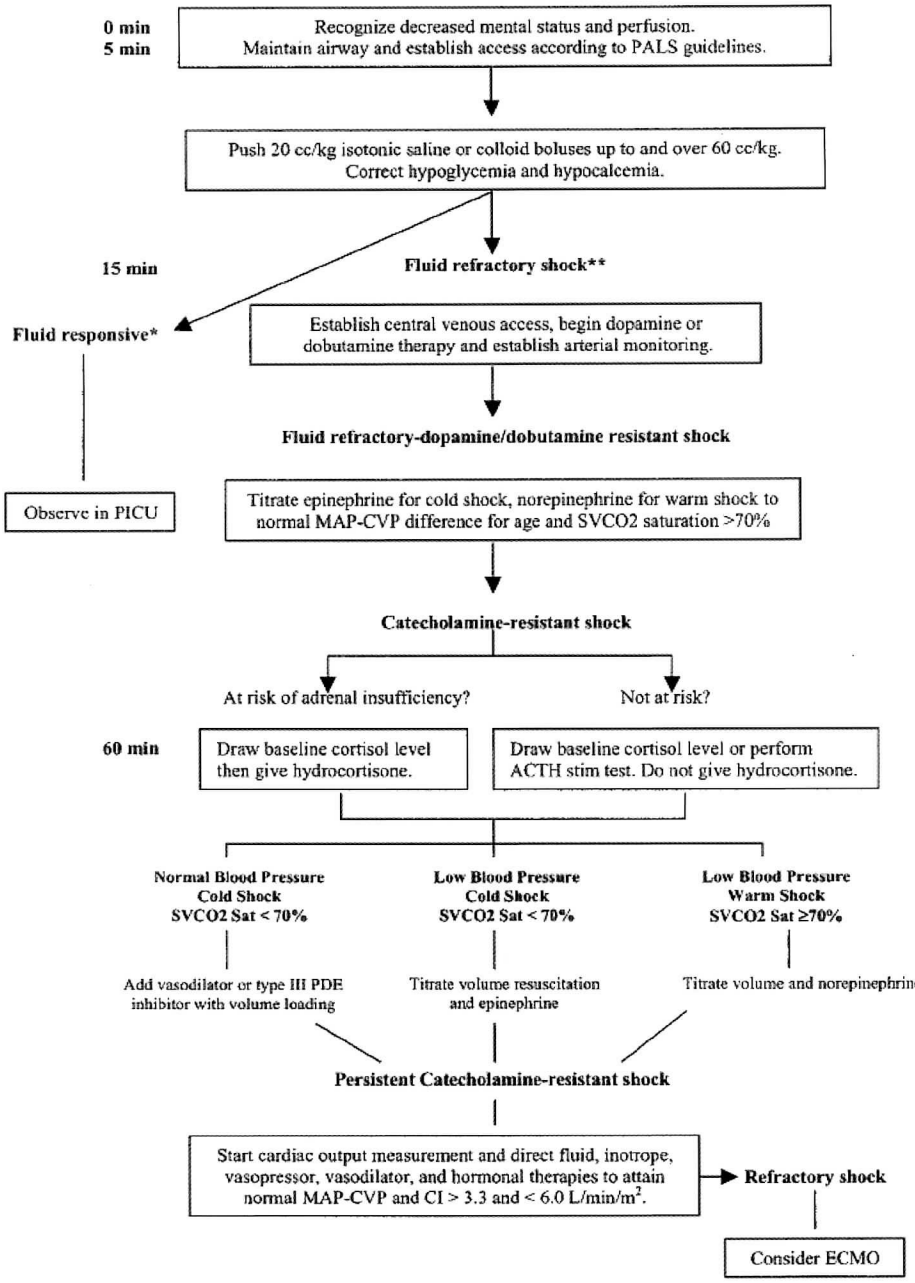
- Catecholamine resistance
- Suspected or proven adrenal insufficiency
  - At Risk:
    - Septic shock and purpura
    - Chronic steroid use
    - Pituitary or adrenal pathology
- ***1-2 mg/kg*** *for* ***stress*** *coverage*
- ***50 mg/kg*** *for empiric therapy for* ***shock***

Differential Diagnosis Of Septic shock

Clinical Signs	**Hypovolemic Shock**	Distributive Shock	Cardiogenic Shock
Respiratory Rate	⇑	⇑ or ⇑⇑	⇑⇑
Respiratory Effort	Normal	Normal to ⇑	⇑⇑
Heart Rate	⇑	⇑ to ⇑⇑	⇑⇑
Pulse Quality	Thready	Early-bounding Late-thready	Thready
Pulse Pressure	Narrow	Widened	Narrow
Skin Perfusion	Pink, cool distally, nl or prolonged CR	Pink, often warm early, nl to long CR	Mottled gray or blue, cool to cold, prolonged CR
Level of Consciousness	Usually normal unless severe	Lethargic or confused	Lethargic to coma
Urine Output	Decreased	Decreased	Markedly decreased
Stroke Volume	Low	Normal to increased	Markedly decreased
Preload	Low	Low	Often high
Afterload	High	Low	High
Acidosis	Mild to moderate	Mild to marked	Moderate to marked

*adapted from PALS Provider Manual, AAP, 2002

Figure . Resuscitation of pediatric septic shock.



Dellinger, R.P. et.al. Surviving Sepsis Campaign guidelines for management of severe sepsis and septic shock. Critical Care Medicine, 32(3), March 2004, 858-73

ACUTE UPPER AIRWAY OBSTRUCTION

Most common presentation is sudden choking or stridor. Stridor could be inspiratory only (indicates extrathoracic obstruction) or could be both inspiratory and expiratory (indicates intrathoracic obstruction). Approach to a choking child has already been discussed. In the initial assessment don't touch the child, leave her in mother's lap. If he is cyanosed give the oxygen mask in mother's hand and ask her to hold it on patient's face.

Look for: Cyanosis, neck position, drooling

How the voice has been affected? Inability to produce sound is a bad sign.

Is he very sick looking, lethargic, agitated & irritable?

Feel: If he is febrile.

Auscultate:

If unequal breathe sound or localized ronchi, think of foreign body. Don't go for blood gas immediately but it is important to see oxygen saturation by pulse oxymeter, if it is available. Whatever you do, explain to the child. Do not attempt to see the throat or to do a lateral neck X-ray in the emergency department if you think the child is very sick. Take him to PICU or OT and call anaesthetist and otolarygologist before throat examination and portable X-ray. Be ready for intubation and tracheostomy.

Etiology: Remember the etiology by letters **A, B, C, D, E, F, G, H**

Asking a few questions you can qualify the etiology easily.

A: Angioneurotic edema: Previous history, family history, puffiness. Other allergic manifestations.

Abscess of the retropharyngeal space: History of pencil injury through, oral cavity, patient febrile, toxic, submandibular swelling, neck extension, throat examination may reveal bulged posterior pharyngeal wall.

B: Bacterial tracheitis: Stridor inspiratory & expiratory, onset over days, patient toxic, highly febrile

C: Croup (viral): Common condition, upper respiratory catarrh, gradual onset, seasonal - may be cluster of cases, stridor louder, child not toxic, fever low grade.

D: Diphtheria: Rare now a days, ask about vaccination. Examination of throat commonly reveals the greyish membrane.

E: Epiglottitis: Acute Onset, high fever, child toxic, drooling, stridor fainter, but retraction profound, child may be cyanosed. Examine the *throat* in PICU (Paediatric Intensive Care Unit) or OT. Uvula looks cherry red.

F: Foreign Body: Afebrile, onset with sudden choking.

G: Growth: Rapidly growing tumor in neck and superior mediastinum (rare).

H: Hypocalcemia: Look for signs of rickets, or hypoparathyroidism.

Management: Will depend on your assessment and underlying cause.

- If very sick, patient is to be taken to PICU or operation theater (OT) where throat examination should be followed by intubation.
- Bronchoscopy & tracheostomy might be needed and it's preferable to include in the team: anesthetist, thoracic & ENT surgeons. After air way

is secured - blood culture, throat swab & tracheal aspirate cultures are to be taken. Antibiotics should be appropriate to cover haemophyllus influenzae for acute epiglottitis we prefer 3rd generation cephalosporin - cefotaxime or ceftriaxone. In suspected bacterial tracheitis antibiotic should cover staph. aureus, pseudomonas as well as haemophillus influenzae. Retropharyngeal abscess should be drained surgically in addition to antibiotics.

- For angioedema: antihistamines IV or IM, adrenaline - IM or SC, corticosteroids IV.

COMATOSE CHILD

Remember causes of coma by letters A, E, I, O, U (vowels) + D, T, M & H.

A: Apoplexy: Synonymous with stroke or intracranial hemorrhage
E: Epilepsy, encephalitis
I: Injury to head, intoxication
O: Opium and other poisoning
U: Uremia
D: Diabètes mellitus
T: Tumour
M: Meningitis, malaria & metabolic e.g., hypoglycemia, etc.
H: Hepatic failure

Initial Assessment:

Is airway patent with sufficient ventilation? Is circulation adequate?
Is intracranial pressure elevated? Is there a focal sign?
Is there a remediable metabolic problem (blood gas & glucose)?

Important Examination Points:

Vital Signs:

Bradycardia, hypertension, Cheyne-Stokes respiration indicates raised intracranial pressure. Look for facial asymmetry, squint, see reflexes, and the fundus. Examine pupils for size and reaction to light.

Glasgow Coma Scale:

1. Eye opening (scores 1 to 4)
2. Verbal response (scores 1 to 5)
3. Motor response (scores 1 to 6)

Minimum score 3 X 1 = 3, maximum 4 + 5 + 6 = 15

Glasgow Coma Scale: Modified for children

Eye Opening			
Score	> 1 year	< 1 year	
(4)	Spontaneously	Spontaneously	
(3)	To verbal command	To shout	
(2)	To pain	To pain	
(1)	No response	No response	
Best Motor Response			
Score	> 1 year	< 1 year	
(6)	Obeys	Spontaneous purposeful movement	
(5)	Localizes pain	Localizes pain	
(4)	Flexion withdrawal	Flexion withdrawal	
(3)	Decorticate rigidity	Decorticate rigidity	
(2)	Decerebrate rigidity	Decerebrate rigidity	
(1)	No response	No response	
Best verbal response			
Score	> 5 years	2-5 years	1-2 years
(5)	Oriented & converses	Appropriate words	Smiles, coos appropriately
(4)	Disoriented & converses	Inappropriate words	Cries, consolable
(3)	Inappropriate words	Persistent cries or screams	Persistent crying
(2)	Incomprehensible sounds	Grunts	agitated, restless
(1)	No response	No response	No response

Glasgow Coma Scale is a good way to monitor level of coma.
Score less than 7 has usually bad prognosis.

Investigations:

Blood sugar, urea, creatinine, electrolytes
Arterial blood gas, ammonia, liver functions, blood count, malarial parasites, blood CS, toxicology screening - (blood, urine & gastric washings)
X-ray Skull, US, CT scan & MRI of brain
Urine for sugar and ketones
LP is contraindicated for raised intracranial pressure - defer until CT scan
CSF Analysis & CS, viral study - for meningitis & encephalitis
EEG, blood and urine for metabolic screening.

Management:

1. Airway: If needed intubate & ventilate, give oxygen, prevent aspiration by semiprone position.
2. Circulation: IV or intraosseous line, maintain blood glucose, electrolytes, restrict fluid therapy.
3. Recognize & treat raised intracranial pressure.
4. Prevent bed sores, exposure keratitis.
5. Balance intake and output, provide care of bladder & bowel.
6. Adequate nutritional management.
7. Find out and treat specific cause.

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CARDIOVASCULAR SYSTEM

EXAMINATION:

- **General comments:**

  - Ill looking or well, central cyanosis (is seen in the tongue), pallor, signs of respiratory distress (tachypnoea, intercostal, subcostal, suprasternal recession, flaring of ala nasi), any connection of oxygen, IV lines.
  - General health (failure to thrive or well nourished).
  - Dysmorphic features (are these features fitting to a known syndrome).

- **Hands:**

  - Nails for clubbing and splinter hemorrhage.
  - Palmer creases for pallor.
  - Look for other signs: tuberous and tendon xanthoma over bony prominence, Osler's nodes at palmer surface of finger tip.

- **Arterial pulse:**

  - Check that both brachial pulses are present and equal in volume.
  - Check for femoral pulses (decreased or absent, suggest aortic coarctation).
  - Check for rate (for at least 30 seconds)
  - Check for rhythm (regular or irregular)
  - Check for volume (large volume suggests PDA, aortic regurgitation).
  - Check for the character (collapsing pulse, slow rising pulse, pulsus pardoxus, and pulsus alternans).

- **Blood pressure (Very important):**

  - For children is measured by sphygmomanometer.
  - For infant is measured by Doppler ultrasound, from all limbs.

- **Check for edema in the lower limbs and sacral region**

Examination of Precordium

- **Inspection:**
  - Asymmetry: bulge of left chest due to cardiomegaly.
  - Other deformities (pectus excavatum, pectus carinatum and Harrison sulcus).
  - Scars: median sternotomy scar, left and right anterolateral scars.
  - Visible pulsation e.g. pulsation at neck in aortic regurgitation, apex beat.
  - Assessment of Jugular venous pressure (JVP) in older children with heart failure.

- **Palpation:**

  - Apex beat (palpable cardiac impulse at the outer most and lower most position, is located normally at 4th intercostal space in the mid-clavicular line): Localize the site properly and comment on quality (heaving or tapping).
  - Parasternal heave: feel over the left parasternal area with the heel of hand for parasternal heave, which indicates right ventricular hypertrophy.
  - Thrills (palpable murmur): Localize the site by palpating all areas including suprasternal notch left subclavicular region and neck. The accompanying murmur is by definition at least 4/6 in intensity
  - Palpable 2nd heart sound: Over pulmonary area, this will be the pulmonary component and reflects pulmonary hypertension.
  - Palpate the abdomen at the end of palpation of precordium for:
    - The liver.
    - The Spleen, if you suspect infective endocarditis by other signs.

- **Auscultation:**

  - Use both bell and diaphragm, the bell for low pitched sound (diastolic murmur at the apex and heart sounds).
  - Palpate right brachial artery with your left hand during auscultation, this will help you to time the murmurs

Follow this order:

  - Apex (Mitral area), if there is murmur check for radiation to left axilla.
  - 4th, 5th intercostal space at left sternal edge (Tricuspid area).
  - 2nd intercostal space at left sternal edge (Pulmonary area).
  - 2nd intercostal space at right sternal edge (Aortic area), if there is systolic murmur listen over the carotid for radiation of the murmur.
  - Left subclavicular area for P.D.A. murmur.
  - Listen at the 4th intercostal space mid-clavicular line on right side to exclude dextrocardia.
  - Listen at the back between scapulae for the murmur of coarctation of aorta and below left scapulae for PDA murmur.
  - If you are suspecting innocent murmur or mitral valve prolapse, listen to the murmur both on sitting as well as supine position.
  - For older child listen again along left sternal edge at the level of 3rd and 4thintercostal space as he breaths out and leans forward for murmur of aortic regurgitation. The murmur of mitral stenosis is heard more clearly if child lies in supine position and turns towards left.

LISTEN FOR THE FOLLOWING:

1) Normal *heart sounds:*
   - 1st heart sound for intensity (best at apex).
   - 2nd heart sound (best at pulmonary area) for intensity and splitting.

2) Added *sounds* (all best heard at apex with the bell of stethoscope except ejection click).
   - 3rd heart sound can be normal finding in children, heard early in diastole.
   - 4th heart sound heard late in diastole.
   - Opening snap after 2nd heart sound in mitral stenosis.
   - Click: midsystolic in mitral valve prolapse at apex, early systolic in aortic and pulmonary valve stenosis best heard at mid left sternal border and at the corresponding areas.

3) Murmur:
   - Location (maximum intensity).
   - Timing (systolic, diastolic or both).
   - Duration (e.g.: pansystolic, ejection systolic, early diastolic).
   - Intensity (grading from 1 to 6, if there is thrill the murmur is more than 3/6 in intensity).
   - Radiation.
   - Quality and pitch (rumbling, blowing, harsh, low pitch, high pitch).
   - Change with posture or phases of respiration (e.g.: venous hum disappear on lying down).

INNOCENT MURMUR

70% of children have innocent murmur at sometime.

Clinical feature:

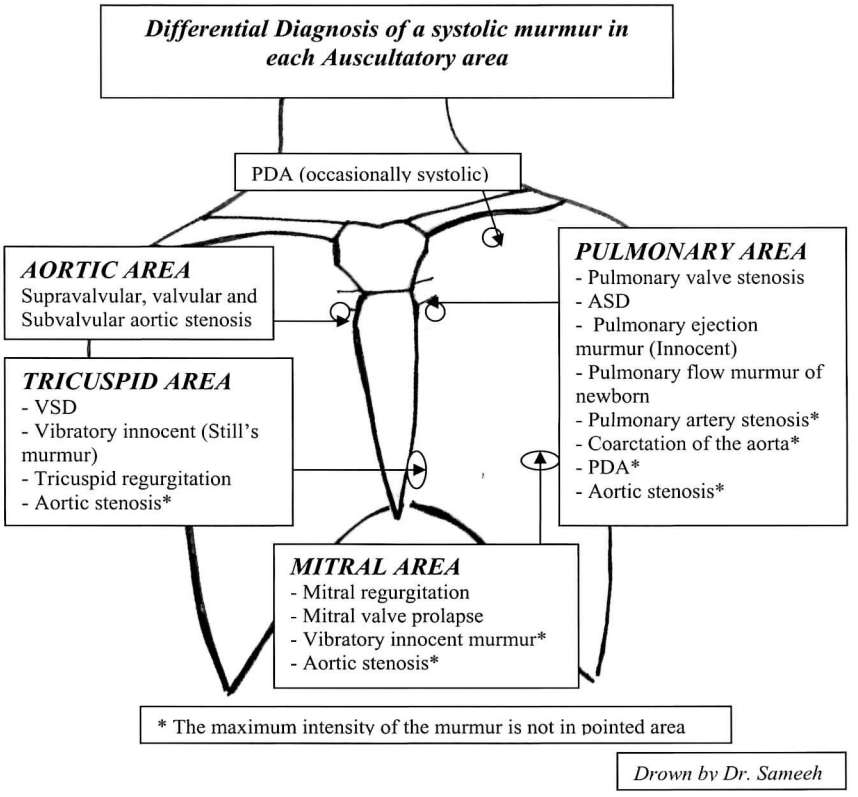
First:
   - The history or examination should not suggest an organic cause.
   - No relevant symptoms.
   - No cyanosis.
   - No signs of heart failure.
   - No abnormality on examination: normal pulses, heart sounds. and cardiac impulse.
   - No thrill.

Second:
   - Innocent murmurs are limited to systole (except venous hum), short, not louder than grade 2/6, no radiation and may vary with posture.

Finally:
   - Normal chest X-ray, ECG and Echocardiography.

Types of innocent murmur:
- Still's murmur:
   - It is usually low-pitched and vibratory in character.
   - Best heard medial to the apex and at the lower part of the left sternal edge.
   - It may decrease in intensity when the child stands up.
   - Differential diagnosis: very small VSD and mitral valve prolapse.

- Basal ejection murmurs (pulmonary and aortic flow murmur):
  - Are soft, ejection systolic murmur.
  - Best heard at the base of the heart either in pulmonary or aortic area,
  - The murmurs are due to increase flow of blood across these valves, more prominent with exercise, fever and anemia.
  - Occasionally pulmonary flow murmur is found in the newborn for 3 - 6 months.
  - Differential diagnosis:
    - ASD.
    - Bicuspid aortic valves with mild aortic stenosis.
    - Mild pulmonary valve stenosis.
    - Hypertrophic cardiomyopathy.
- Venous hum
  - Is blowing, continuous murmur.
  - Best heard at the base of the heart just below the clavicles especially on the right side and while the child is standing or sitting.
  - It varies both with respiration and the position of the head (disappears when the child lies down or when he turns his head to the same side)
  - Differential diagnosis: P.D.A.



NORMAL HEART RATE AT REST

AGE	NORMAL RANGE *(Beat / minute)*	AVERAGE
0 – 1 Month	70 -1 90	125
1 - 11 Months	80 - 160	120
2 years	80 - 130	110
4 years	80 - 120	100
6 years	75 - 115	100
6-12 years	70 - 110	90

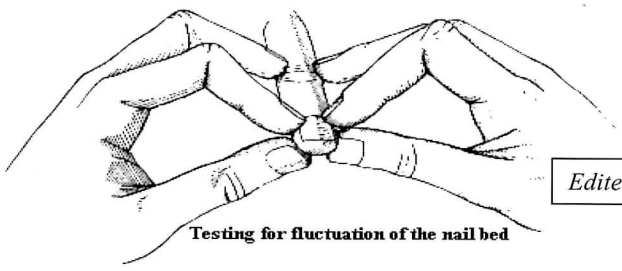
Causes of tachycardia:

- Sinus tachycardia
  - Hyperdynamic circulation: Exercise, anxiety, anemia, pyrexia, thyrotoxicosis.
  - Heart failure, shock, constrictive pericarditis.
  - Drugs e.g. aminophylline, salbutamol, adrenaline.
- Supraventricular (atrial or nodal/AV junctional) tachycardia
- Atrial flutter, atrial fibrillation.
- Ventricular tachycardia

Causes of bradycardia:

- Sinus bradycardia
  - Congenital
  - Athletes
  - Drugs e.g. beta-blockers, digoxin.
  - Hypothyroidism, hypothermia, increased intracranial pressure, hyperkalemia.
- Complete heart block
  Maternal SLE, Structural heart disease, Surgical trauma, Antiarrhythmic drugs.
- Second degree AV block
  Rheumatic fever, Ebstein's anomaly, digitalis toxicity

Finger clubbing



Testing for fluctuation of the nail bed

Features: - Increase fluctuation of the nail bed.
- Loss of the angle between the nail and nail bed.
- Increase curvature in both dimensions.

- Causes of clubbing:
  - Cardiac causes:
    - Cyanotic congenital heart disease, e.g. tetralogy of fallot.
    - Infective endocarditis.
  - Pulmonary causes:
    - Chronic suppurative lung disease.
    - (Bronchiectasis, Pulmonary abscess, empyema).
    - Cystic fibrosis.
    - Fibrosing alveolitis.
  - GIT causes:
    - Biliary cirrhosis, biliary atresia, ulcerative colitis, crohn's disease.
  - Other rare causes:
    - Thyrotoxicosis - Malignant neoplasm of lung.
    - Familial and idiopathic.

Chest scar

1. Left anterolateral thoracotomy scar:
   - *Cardiac operation:*
     - PDA ligation (normal pulse in left arm)
     - Pulmonary artery banding (normal pulse in left arm).
     - Repair for coarctation of aorta (usually normal pulse in left arm).
     - Blalock taussing shunt: Classical (weak pulse in left arm).
     - Blalock taussing shunt: Modified (normal pulse in left arm).
   - *Non cardiac:*
     - Lobectomy of left lung.
     - Diaphragmatic eventration.
2. Right anterolateral thoracotomy scar:
   - *Cardiac operation:*
     - Blalock taussing shunt: Modified or classical.
     - Glenn procedure.
   - *Non cardiac:*
     - Lobectomy of right lung.
     - Diaphragmatic eventration.
     - Esophageal atresia repair.
3. Median sternotomy scar (Central sternotomy):
   - Open-heart surgeries e.g. repair of septal defect or valve replacement.

Cyanosis

Peripheral cyanosis:

It is not uncommon in neonate and young infants; it may merely signify that child is feeling cold.

Causes:

1. Physiological -- cold
2. Reduced cardiac output
3. Peripheral vascular disease e.g.: Polyarteritis nodosa

Central cyanosis:

Best seen in the tongue, it may be obvious at rest or only after exertion such as feeding or crying. It is detectable if there is deoxygenated hemoglobin of more than 5 gm/100 ml. Blood, corresponding to an arterial saturation of 75%.

Causes:

Cardiac: Most of them begin with the letter "T"

* Cyanotic congenital heart disease presenting in neonate
  - Transposition of the great arteries.
  - Tricuspid atresia.
  - Total anomalous pulmonary venous drainage.
  - Truncus arteriosus.
  - Tricuspid regurgitation with Ebstein's anomaly.
  - Pulmonary atresia.
  - Complex congenital heart disease e.g. single ventricle with critical pulmonary stenosis.
* Cyanotic congenital heart disease presenting after neonatal period:
  - Tetralogy of Fallot.

Pulmonary causes:

* Neonatal period:
  - Respiratory distress syndrome.
  - Aspiration pneumonia (meconium, milk).
  - Congenital pneumonia.
  - Diaphragmatic hermia
  - Lung hypoplasia
* After neonatal period:
  - Pneumonia
  - Bronchiolitis
  - Bronchial asthma
  - Cystic fibrosis
  - Bronchiectasis

Nervous system and neuromuscular causes:

a. Central nervous system lesions, e.g. birth asphyxia, head trauma.
b. Central nervous system depression by drugs, e.g. phenobarbitone.
c. Intercostal muscle weakness, e.g. spinal muscular atrophy.
d. Diaphragmatic weakness, e.g. phrenic nerve palsy.
e. Other causes:
   i. Persistent pulmonary hypertension of the newborn (persistent fetal circulation) the oxygen saturation of the right arm (preductal) is more than the oxygen saturation from the legs (postductal).
   ii. Methemoglobinemia (very rare). Pao_2 is normal but the saturation is low.
   iii. Polycythemia.

iv. Non cyanotic heart disease with heart failure (cyanosis is mainly due to pulmonary edema) e.g. hypoplastic left heart, Intra pulmonary arteriovenous malformation with right to left shunt.

N.B.:

- **The hyperoxic test:** This test helps to differentiate cyanotic congenital heart disease from lung disease and nervous system disorder. An arterial blood for gas analysis should be taken from the right arm if possible (preductal circulation) and then should be repeated after administration of 100% oxygen for 10 minutes. Oxygen is best delivered via a head box, mask, endotracheal tube.

Interpretation of hyperoxic test

After performing the hyperoxic test if:

1. PaO_2 is < 100 mmHg the cause is most likely cyanotic heart disease
   - Transposition of great arteries.
   - Condition with decreased pulmonary blood flow: Pulmonary stenosis, pulmonary atresia, tricuspid atresia, and Ebstein's anomaly with pulmonary stenosis total anomalous pulmonary venous drainage (obstructive type). Prostaglandin therapy is useful in these cases.
2. PaO_2 is > 100 mmHg, especially > 150 mmHg the most likely causes are:
   - Primary lung disease.
   - Nervous system and neuromuscular disorder.

PRESENTATION AND MANAGEMENT OF HEART DISEASES IN THE NEONATAL PERIOD

Severe heart disease in the newborn, may manifest as:

- **Cyanosis**:
  e.g. Transposition of great arteries
  Pulmonary atresia
  Tricuspid atresia
- **Congestive heart failure**
  e.g. Complete atrioventricular septal defect
- **Shock**
  e.g. Severe coarctation of aorta
  Hypoplastic left heart syndrome
- **Cyanosis with distress (cyanosis and heart failure)**
  e.g. Transposition of great arteries.
  Total anomalous pulmonary venous drainage.
  Complex congenital heart disease

- **Management of newborn with serious heart disease**

A) Management in primary health center or peripheral hospital:

- Any neonate with cyanotic heart disease or heart failure should be transferred immediately to specialized unit for investigation and treatment and require urgent consultation with paediatric cardiologist.
- Care should be taken to keep the infant warm, in oxygen and under close observation.
- 30-40% Oxygen in the inspired air (1liter/min by nasal prong or nasal catheter) is sufficient to improve arterial Pao_2 (if arterial PaO_2 is low due to pulmonary edema or chest infection). High oxygen concentration is not advised because it may facilitate ductus arteriosus closure in the ductus dependent lesion and increases heart failure in congenital heart diseases with left to right shunt such as VSD.
- Intravenous fluid (5-10% dextrose in 0.225% normal saline) should be started. Regular fluid maintenance is advised to cyanotic neonate to avoid dehydration and risk of thrombosis; however for neonates with heart failure restriction of the fluid to 70% is necessary.
- Blood gases, serum glucose and calcium should be checked if possible.
- In the presence of severe congestive heart failure, intravenous furosemide should be given. Avoid diuretic therapy for the baby with severe cyanosis without significant heart failure.
- When the cardiac center is far from the referring doctor and in the presence of ductus-dependent cardiac lesion, prostaglandin E_1 or E_2 should be started in appropriate dosage after discussion with the paediatric cardiologist.

B) Initial management of a neonate with heart disease in medical center (Big hospital):

- The infant should be kept warm in an incubator with 30-40% oxygen.
- IV fluid 5-10% dextrose in 0.225% normal saline.
- Basic investigations include blood glucose, serum calcium, magnesium, other electrolytes, complete blood count, blood gases and infection screen.
- Chest X-ray and ECG
- Hyperoxic test for cyanotic neonate.
- Monitor oxygen saturation or Arterial PO_2 from right arm and either leg
- Measurement of blood pressure in both arms and legs.
- Contact paediatric cardiologist for consultation regarding further management or for possible transfer to cardiac center.
- Prostaglandin infusion for ductus dependent lesion after discussion with paediatric cardiologist.
- Anti-failure drugs (diuretic, digoxin) for a cardiac lesion presenting with heart failure.
- Correction of metabolic acidosis with sodium bicarbonate.
- Inotropic support with dopamine/ dobutamine for cardiac lesion presenting with shock or acute heart failure.
- Other measures for heart failure depends on severity including:

- Elevation of the head end of the bed to 40 degree (may help).
- Intubation and ventilation with positive end expiratory pressure.
- Morphine sulfate subcutaneously or IV 0.1 mg/kg.

Ductus dependent lesions:

A number of cardiac lesions exist in which either the pulmonary or the systemic blood flow is greatly ductus dependent, these lesions will not manifest until after birth, at the time of ductal closure. Prostaglandin administered prior to anatomical ductal closure raises the systemic arterial oxygen saturation, improves perfusion, stabilizes the infant and gives time for cardiac catheterization or surgical intervention.

Ductus - dependent cardiac lesions are divided into three categories.

1. *Cyanotic lesions with obstruction to pulmonary blood flow*:
   a) Pulmonary atresia.
   b) Critical pulmonary valve stenosis.
   c) Tricuspid atresia with critical pulmonary valve stenosis.
   d) Complex cyanotic cardiac lesion with critical pulmonary stenosis.
2. *Acyanotic lesions with obstruction to systemic blood flow*:
   a) Pre or juxta - ductal coarctation of aorta.
   b) Critical aortic stenosis.
   c) Hypoplastic left heart syndrome.
   d) Interrupted aortic arch.
3. *Transposition of great arteries*.

Prostaglandin infusion should be used in intensive care unit, because of serious side effects (apnea, hypotension and jitteriness). The blood pressure and respiratory activity should be monitored carefully.

Side-effects of Prostaglandin

Immediate:

Cardiovascular	: Hypotension, cutaneous vasodilatation & rhythm disturbance.
Central nervous	: Pyrexia, jitteriness &muscle twitching.
Respiratory	: Respiratory depression & apnea.
G.I.T	: Diarrhea & necrotizing enterocolitis.
0thers	: Hematological (abnormal platelet function & D.I.C). Metabolic (hypokalemia). Infection.
Long term	: Cortical hyperstosis, friability of PDA & damage to pulmonary arterial smooth muscle.

The decision to infuse PGE for a neonate with cyanotic heart lesion should be made in consultation with a paediatric cardiologist, as there are risks in giving PGE to infants with total anomalous venous return or persistent pulmonary hypertension.

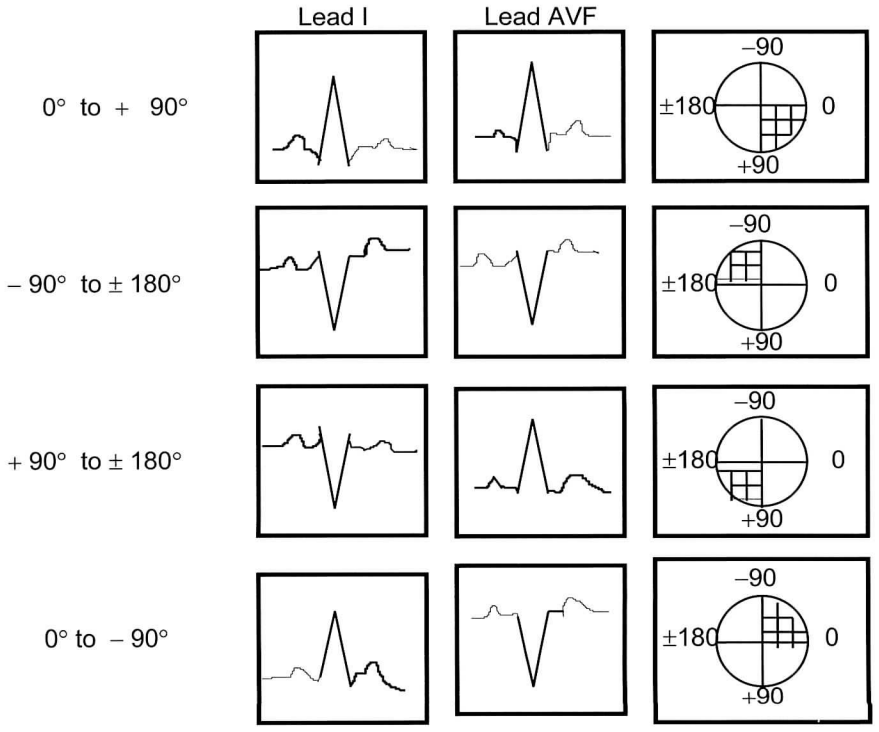
Chest X-ray findings characteristic for certain cyanotic heart diseases:

- Boot-shaped heart: Tetralogy of Fallot.
- Egg-shaped heart: Transposition of great arteries.
- Snowman (figure of 8): Total anomalous pulmonary venous drainage.

ELECTROCARDIOGRAM (ECG)

A) Rate is age dependent (Newborn: 100-160, infant: 80-140, toddler: 75-120, child over 6 years 70-100). Each small division on ECG paper is 0.04 seconds, each large division equals 0.2 second. When the heart is fast, count the RR cycles in 6 large divisions (1.2 seconds) and multiply them by 50. When the heart rate is slow, count the number of large divisions between two R waves and divide into 300 or count the number of small division and divide by 1500.

B) The QRS axis is age specific, it also changes with ventricular hypertrophy or conduction defects. Locate a quadrant, depending on the deflections of the QRS complexes on leads I and AVF.



The mean QRS axis according to age is as follows:

Newborn	+125 degrees
1 month	+90 degrees
3 years	+60 degrees
Adult	+50 degrees

C. Rhythm should be assessed for regularity and voltage. Voltage may be decreased in presence of myocarditis or pericardial effusion.

D. P wave implies atrial activity it is seen best in leads II, III and AVF normal width is 0.08 0.10 second.
   - Left atrial hypertrophy (P-mitral): Prolongation of the P duration, greater than 0.10 second in any lead, most often in lead II. A broad and notched P wave in lead II and III is characteristic and P wave is biphasic in V_1 with a negative prolonged, terminal segment (> 0.04 second duration and deflection > 1 mm)
   - Right atrial hypertrophy (P - pulmonale): Peaked P wave, 3 mm or greater, in any lead, most often in lead II.

E. PR interval (from the beginning of P wave to the beginning of the Q wave) is age dependent and ranges from 0.10-0.20 second.
   - Prolongation of PR interval indicates first degree AV block that occurs with myocarditis (rheumatic, viral or diphtheric), drugs (digitalis, propranolol, verapamil), ischemia or severe hypoxia and hyperkalemia.
   - Short PR interval: WPW syndrome and glycogen storage disease type II (Pompe's disease).
   - Variable PR intervals are seen in wandering atrial pacemaker and type I second degree AV block (Wenckebach phenomena)

F. QRS complex (from the beginning of the Q wave to the end of the S wave) is normally less than 0.10 second. Widened QRS complex indicates an abnormal delay of the impulse through the ventricle which is seen in:
   - Bundle Branch Block, WPW syndrome, Ventricular pacemaker and Arrhythmias of ventricular origin e.g. in severe hyperkalemia. Large QRS voltage occurs with ventricular hypertrophy.

   Left Ventricular Hypertrophy:
   - R in V_6 + S in V_1 greater than 60 mm.
   - R in V_6 greater than 25 mm (> 21 mm under 1 year).
   - S in V_1 greater than 30 mm (> 20 mm under 1 year).
   - An increase in the R/S ratio in V_5, V_6.
   - S in V_1 greater than twice R in V_5.
   - Left axis deviation for the patient's age (LAD).

   Right Ventricular Hypertrophy:
   - R in V_1 greater than 20 mm (after 1 month).
   - S in V_6 greater than 6 mm (after 1 month).
   - An increase in the R/S ratio in V_1, V_2.
   - Upright T wave in V_3 R or V_1 after 3 days.
   - QR pattern in V_1, V_3 R.
   - Right axis deviation for the patient's age (RAD).

Combined ventricular hypertrophy:

- Large equiphasic QRS complexes in mid - precordial leads V_3 - V_4 greater than 70 mm.
- RVH and S in V_1 or R in V_6 greater than mean for age.
- LVH and R in V_1 or S in V_6 greater than mean for age.

RVH in Newborn:

1. Pure R wave in V_1 greater than 10 mm.
2. R in V_1 greater than 25 mm or R in AVR greater than 8 mm.
3. QR pattern in V_1 (can occur in 10% of normal newborn).
4. Upright T in V_1 after 3 days of age.
5. RAD greater than + 180 degree.

G) ST segment abnormalities (elevation or depression) occur with myocardial ischemia, pericarditis, digitalis, myocarditis, severe ventricular hypertrophy hypokalemia and hyperkalemia.

H) Tall T wave is seen in hyperkalemia and LVH. Flat or low T-wave is seen in hypokalemia, myocardial ischemia, hypo or hyperglycemia, myocarditis, pericarditis, hypothyroidism and digoxin.

I) QT segment duration (from the beginning of Q wave up to the end of the T wave) is rate dependent, therefore must be corrected by Bazett's formula:

$$\text{Corrected QT interval} = \frac{\text{QT measured}}{\text{Sqr Rt of RR interval}}$$

Normal: 0-6 months < 0.49 sec, > 6 months < 0.425 sec
It is prolonged with antiarrhythmic agents, tricyclic antidepressants, phenothiazines, hypokalemia, hypocalcemia, hypomagnesaemia, hypothermia, mitral valve prolapse and congenital long QT syndromes.
Short QT interval is seen in hypercalcemia and digitalis toxicity

ECG findings characteristic for certain congenital heart malformations:

A) L.A.D. (Left axis deviation):
- ASD (Premium).
- Complete AVSD.
- Tricuspid Atresia.

B) W.P.W. (Wolff-Parkinson-White) syndrome:
- Ebstein's anomaly.
- L - T.G.A.

C) Complete heart block:
- L - T.G.A.
- Polysplenia.

CONGENITAL MALFORMATION SYNDROMES ASSOCIATED WITH CONGENITAL HEART DISEASE

SYNDROME	THE COMMON ASSOCIATED HEART DEFECT
DOWN'S SYNDROME (Trisomy 21)	AVSD, VSD., ASD
EDWARD'S SYNDROME (Trisomy 18)	VSD. ASD. AORTIC COARCTATION
PATAU'S SYNDROME (Trisomy 13)	VSD. P.D.A., DEXTROCARDIA, AORTIC COARCTATION
CRI DU CHAT SYNDROME(Deletion short arm of 5)	VSD., PDA, ASD
TURNER'S SYNDROME (45X0)	COARCTATION OF AORTA, AORTIC STENOSIS.
NOONAN'S SYNDROME	PULMONARY VALVE STENOSIS, CARDIOMYOPATHY, ASD
WILLIAM'S SYNDROME	SUPRAVALVULAR AORTIC STENOSIS, PERIPHERAL PULMONARY STENOSIS
FRAGILE X SYNDROME	MITRAL PROLAPSE, AORTIC ROOT DILATATION
HOLT-ORAM SYNDROME	ASD., VSD.
FETAL ALCOHOL SYNDROME	ASD., VSD., TETRALOGY OF FALLOT
CONGENITAL RUBELLA SYNDROME	PERIPHERAL PULMONARY STENOSIS P.D.A.
MARFAN'S SYNDROME	AORTIC REGURGITATION, MITRAL PROLAPSE, DISSECTING AORTIC ANEURYSM.
INFANT OF DIABETIC MOTHER	HYPERTROPHIC CARDIOMYOPATHY, VSD, TAG., TOF
THROMBOCYTOPENIA AND ABSENT RADIUS (TAR)	ASD, TOF
DIGEORGE SEQUENCE	AORTIC ARCH ANOMALIES, TETRALOGY OF FALLOT
CHARGE	VENTRICULAR, ATRIOVENTRICULAR AND ATRIAL SEPTAL DEFECTS
ALAGILLE (ARTERIOHEPATIC DYSPLASIA)	PERIPHERAL PULMONARY STENOSIS
DE LANGE'S	TETRALOGY OF FALLOT, VENTRICULAR SEPTAL DEFECT
ASPLENIA	COMPLEX CYANOTIC HEART DISEASE ANOMALOUS VEINS, PULMONARY ATRESIA
POLYSPLENIA	COMPLEXE ACYANOTIC LESIONS, AZYGOUS CONTINUATION
HYDANTOIN	ATRIAL OR VENTRICULAR SEPTAL DEFECT, COARCTATION
VALPROATE EFFECTS	COARCTATION, HYPOPLASTIC LEFT HEART

ACYANOTIC LESION WITH LEFT TO RIGHT SHUNT

CARDIAC LESION	HEART SOUND	MURMURS (MAXIMUM INTENSITY)
VSD	Normal, occasionally loud second sound (large VSD)	Loud, harsh pansystolic murmur at left sternal edge in the 3rd & 4th intercostal space. Occasionally short systolic (very small VSD) Mid diastolic mitral flow murmur is associated with large defect.
ASD	Wide fixed splitting of second sound	Ejection systolic murmur at pulmonary area, tricuspid diastolic flow murmur at left sternal edge with large defect. In ASD premium additional pansystolic murmur may be heard in the apex due to cleft in mitral valve leaflet.
PDA	Normal (loud S2 in large shunt)	Continuous murmur below left clavicle, radiates through to the back (machinery). In large shunt, murmur is systolic only (due to pulmonary hypertension), and is associated with mitral mid diastolic flow murmur at the apex.

CYANOTIC HEART DISEASE

CARDIAC LESION	HEART SOUND	MURMUR
Tetralogy of Fallot	Single second heart sound	Ejection systolic murmur in the pulmonary area.
Transposition of the great arteries.	Single second heart sound	Usually no murmur.
Tricuspid atresia.	Single second heart sound	Usually soft systolic Murmur at lower sternal edge.
Pulmonary atresia.	Single second heart sound	Usually no murmur, occasionally pansystolic murmur due to tricuspid regurgitation, rarely PDA continuous murmur.
Total anomalous pulmonary venous drainage.	Wide splitting of second sound, occasionally loud second sound (obstructive type).	Usually no murmur, Occasionally pulmonary flow murmur (ejection systolic at pulmonary area).
Ebstein's Anomaly.	Triple and quadruple Gallops are common.	Rumbling mid diastolic murmur at lower left sternal edge due to tricuspid valve deformity. Systolic murmur suggest associated defect (P.S.)
Truncus arteriosus.	Loud single second sound	Usually a systolic murmur at the left sternal border and a mid diastolic rumble murmur in the apex. Occasionally diastolic murmur of truncal regurgitation is audible.

OBSTRUCTIVE LESION

CARDIAC LESION	HEART SOUND	MURMURS
Pulmonary stenosis	Wide splitting of second sound, single second sound in severe stenosis.	Ejection systolic murmur in the pulmonary area, following ejection systolic click.
Aortic stenosis	1. Narrow splitting of second sound (mild). 2. Single second sound (moderate). 3. Paradoxical splitting of second sound (severe stenosis).	Ejection systolic murmur in aortic area. It radiates to the neck, may be heard also at apex and lower left sternal edge. It may be preceded by ejection systolic click.
Coarctation of aorta	Normal	Short systolic murmur at left sternal edge and between scapulae, occasionally murmur of collaterals over the scapula (continuous).
Hypoplastic left heart syndrome	Normal or single second sound.	Occasionally systolic murmur at left sternal edge.
Pulmonary hypertension	Loud second sound (pulmonary component).	Ejection systolic murmur in the pulmonary area. Early diastolic murmur in the pulmonary area.

VALVULAR LESIONS

CARDIAC LESION	HEART SOUND	MURMUR
Mitral valve prolapse	Normal	Late systolic murmur at apex. It is often preceded by a mid systolic click, more prominent with standing and the valsalva maneuver.
Mitral regurgitation	Wide splitting of the second heart sound occasionally 3rd heart sound.	Blowing pansystolic murmur at apex radiates to left axilla.
Mitral stenosis	Loud 1st heart sound	Low-pitched mild diastolic rumbling murmur at apex proceeded by opening snap.
Aortic regurgitation	Normal	High pitched early diastolic, blowing murmur over left sternal edge. It is best heard with expiration.
Tricuspid regurgitation	Normal or wide splitting of the second sound. Narrow splitting in the presence of the pulmonary hypertension.	Pansystolic murmur at lower part of left sternal edge.
Pulmonary regurgitation	Normal or narrowly splitting second sound.	Early diastolic murmur in the pulmonary area.

HYPERTENSION

Hypertension is defined as an average diastolic or systolic blood pressure exceeding the 95th percentile for age and sex measured on at least three occasions.

CLASSIFICATION OF HYPERTENSION BY AGE AND GROUP

Age Group	*Significant Hypertension (mmHg)*	*Severe Hypertension(mmHg)*
Newborn		
7 Days	SBP≥96	SBP≥106
8-30 Days	SBP≥104	SBP≥110
Infant (< 2 yr)	SBP≥112	SBP≥118
	DBP≥74	DBP≥82
Children		
3 - 5 yr	SBP≥116	SBP≥124
	DBP≥76	DBP≥84
6 - 9 yr	SBP≥122	SBP≥130
	DBP≥78	DBP≥86
10 - 12 yr	SBP≥126	SBP≥134
	DBP≥82	DBP≥90
Adolescent		
13 - 15 yr	SBP≥136	SBP≥144
	DBP≥86	DBP≥92
16 - 18 yr	SBP≥142	SBP≥150
	DBP≥92	DBP≥98

(SBP= Systolic blood pressure -DBP= Diastolic blood pressure)

CAUSES OF HYPERTENSION IN CHILDREN AND ADOLESCENTS

Renal Disease
- Renal artery stenosis

Cardiovascular
- Coarctation of the aorta

Endocrine
- Mineralocorticoid excess
  - Primary hyperaldosteronism
  - 11B - Hydroxylase deficiency
  - 17α - Hydroxylase deficiency
  - Dexamethasone - suppressible hyperaldosteronism
  - Apparent mineralocorticoid excess
- Hyperthyroidism
- Pheochromocytoma
- Hypercalcemia

Tumors
- Neurofibromatosis
- Neurogenic tumors

Others
- Immobilization - induced
- Essential hypertension

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RESPIRATORY SYSTEM

HISTORY:
Please refer to the chapter dealing with the history taking. In addition give particular emphasis to the following points:

- ***Cough***:
  - Character: barking.
  - Duration: acute or chronic (more than 3 weeks)
  - Severity: interfere with sleeping, feeding, and speaking.
  - Painful cough: lesion related to the pleura or ribs (pleurisy, rib fracture)
  - Timing: more at night, seasonal (asthma)
  - Dry or productive: dry in pleurisy, productive in pneumonia.
    Notice character of sputum:
    - Nature: purulent, mucoid, frothy.
    - Quantity: scanty or copious.
    - Color: blood stained (pneumonia, mitral stenosis), greenish (cystic fibrosis), yellowish (pneumonia).
    - Smell: fetid (lung abscess, cystic fibrosis, bronchiectasis)
- ***Difficulty in breathing***
  - At rest or on exertion (during feeding in infant)
- ***Choking during feed or inability to complete feeds.***
- ***Bluish discoloration of the lips (cyanosis)***
- ***Presence of abnormal sounds during breathing***
  - **Wheezing**: may be heard without stethoscope.
  - **Stridor**: "Harsh high pitched sound, heard during or at the end of inspiration"
  - **Snoring**: "Stridor that occurs at sleep"
  - **Grunting**: "Noise produced at the beginning of expiration by a forceful expiration against a partially closed glottis"
  - Rattling**: "Rapid succession of short, sharp sounds due to passage of air in pooled saliva in the throat"**
- ***Other important symptoms:***
  - **Fever**: mention character
  - **Loss of weight**: acute or chronic
  - **Presence of loose fuel smelling stools**. **Chronic diarrhea**
  - Known **chronic diseases**: neuromuscular, cardiopulmonary, immunodeficiency.
  - **Gestational age**, history of ventilation, family history of atopic disorders.

CLINICAL EXAMINATION OF THE RESPIRATORY SYSTEM
General evaluation: Look for -

- General appearance: well or ill looking.
- State of alertness
- Color (pallor, cyanosis)
- Speech ability (reading Qura'an, counting 1-10 in one breath)
- Tachypnea (respiratory rate > expected for age)
- Grunting
- Active ala-nazi

- Audible wheezes
- Stridor
- Snoring
- Built and nutrition
- Note the presence of: Oxygen supply and delivery systems (nebulizer apparatus, spacer devices, and inhaler devices) sputum pot.

Examination of upper limbs: Check for:

- Clubbing: increased rounded appearance of the nails with obliteration of the angle between the nail and its soft tissue base, or a positive Scramroth's sign (obliteration of the diamond shaped space when the nail beds of two corresponding fingers of both hands meet together).
- Peripheral cyanosis.
- Pulse: count and note the character (bounding pulse - CO_2 retention) "count apex beat in children below the age of 3 years".
- Blood pressure: Pulsus paradoxus (> 10 mm Hg fall of systolic blood pressure during inspiration) e.g. severe asthma, constrictive pericarditis.

Examination of the face: Check for:

- Lips: peripheral cyanosis
- Tongue: central cyanosis
- Ala-nazi: active or not

Examination of the Ear, Nose and Throat:

Keep it for the last, but remember to do it. Explain to the patient or use proper restrain as indicated.

- ***Throat:***
  - Use a good source of light and a strong wooden spatula.
  - Don't miss a spontaneous gag to have a clear view of the throat.
  - Don't over-interpret tonsillar enlargement in a gagging child.
- ***Ears:***
  - Use proper size speculum.
  - Gently pull the auricle upward, backward and laterally to make the external auditory canal straight to have a better view of the tympanic membrane. Note: presence of wax or foreign body.
  - Examine the tympanic membrane for: light reflex, color (remember the ear drum may appear reddish in a crying child), bulging or retracted, perforation, discharge and mobility (by a pneumatic device in the otoscope)
- ***Nose:***
  - Use large size speculum and the usual otoscope (if indicated)
  - Look for:
    - Foreign body
    - Foul smell
    - Bleed
  - Color of the nasal mucosa.
  - Presence of discharge and its character
  - Turbinate hypertrophy
  - Polyps
  - Edema of the nasal mucosa (allergy)

EXAMINATION OF THE CHEST:

Examine both *front* and *back* of the chest. Proceed to the back after completion of the examination of the front.

Inspection:

- Respiratory rate (minimum 30 seconds)
- Expose the chest fully and look for:
    - Type of breathing:
        - Abdominal in infants.
        - Thoracic after 4 - 5 years of age
        - *Flat abdomen with reduced movements indicates diaphragmatic hernia*
    - Use of accessory muscles: suprasternal, intercostal and subcostal retractions.
    - Apex pulsation.
    - Scars.
    - Shape of the chest:
        - Pectus Excavatum (funnel shaped chest)
        - Pectus Carinatum (pigeon shaped)
        - Barrel shaped (increased antero-posterior diameter)
        - Harrison Sulcus (indrowing of the lower chest with rib flaring)
        - Chest wall asymmetry and unequal chest movement *(should be observed from the foot end of the bed, with keeping your eye in the same level of chest wall)*
        - Shield-shaped chest: Turner's syndrome
        - Flattening of the hemi-chest, absence of pectoralis muscle i.e., Poland anomaly
        - Others:
            - Rachitic rosary
            - Absent clavicle in cleidocranial dysplasia
            - Supernumerary nipple in renal anomaly

Palpation.

Proceed gently with warm hands.

Note:

- Obvious swelling and tenderness
- Position of trachea: by comparing the gap between the sternal head of the sternomastoid and tracheal margin by your index finger. Normally trachea is slightly deviated to the right. In young children this is not a reliable sign to detect mediastinal shift
- Position of the apex beat. *Remember dextrocardia*
- Tactile vocal fremitus: (in older children) place the palm of the hand on either side of the upper chest and ask the child to say ninety-nine. Feel the difference between sides rather than absolute increase or decrease.
- Chest expansion: (in older children) hook little fingers of both hands in the axillae with thumbs meeting in the mid-line. Ask

the child to take a deep breath and observe which thumb moves the least.

Percussion.

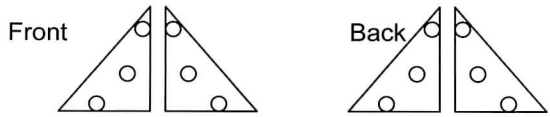
1. Perform very gently
2. Explain to the older patients and the attendants
3. Use:
   A) Pleximeter finger: placed in an intercostal space flush with the chest wall, other fingers kept away from touching the chest wall.
   B) Percussing finger (plexor): middle finger of the dominant hand. Finger should pivot at wrist and not at the elbow, with a gentle blow hitting perpendicularly to pleximeter finger.
4. Percuss at: (corresponding points) from up to down.
5. Mid-clavicular lines, and
6. mid-axillary lines

Note:

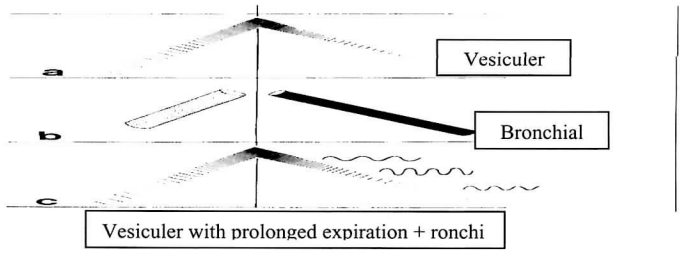
7. apices: percuss on the clavicle directly
8. Area of the liver dullness
9. Area of cardiac dullness
10. Character of resonance: (normal, increased, reduced, absent, stony dullness)

Auscultation.

1. Use a child size stethoscope
2. Be sure that the chest piece is adequately warm
3. Use either the bell or the diaphragm (practice the use of any one).
4. Apply the chest piece firmly to the chest wall to avoid rubbing noises and escape of breath sounds
5. Auscultate the corresponding points in both sides:



1. *Character of air entry*:
   - Normal
   - Reduced
   - Equality on both sides
2. *Character of breath sounds:*



Character of inspiratory and expiratory phase: Note whether,

- Inspiration prolonged
- Expiration prolonged

3. *Presence of added sounds:*
   - Wheezes: continuous, uninterrupted, musical sound
   - Inspiratory: example: croup
   - Expiratory: example: asthma
   - Crackle: discontinuous, interrupted sounds like popping of bubble which may be:
     - Course: i.e., friction rub (rubbing leathery sound)
     - Medium
     - Fine crepitations may also be:
     - Early inspiratory i.e., in obstructive airway disease
     - Late inspiratory i.e., in restrictive airway disease
4. *Vocal resonance:* (in older children)
   As in the tactile vocal fremitus but use chest piece instead of hands. It may be: Increased, Reduced, Absent
5. *Transmitted sounds:*
   May confuse the added sounds, transmitted sounds may disappear by:
   - Clearing the secretion by:
     - Cough - suction - physiotherapy
   - Prior hearing of throaty sounds by naked ear (rattling)
   - Putting the stethoscope by the side of the neck, added sounds increase in intensity

Examination of the back of the chest.

If possible make the child seat and place his hands on his head. Then follow the sequence of:

Inspection: Look for
- Scoliosis and kyphosis
- Scar
- Position of the scapula: one scapula is higher in Sprengel's deformity

Palpation: Look for
- Chest expansion
- Tactile vocal fremitus

Percussion:
- Percuss medial to scapula, liver dullness normally starts adjacent to the spine at the 10th. rib.

Auscultation:
- As in the front of the chest

Examination of the young and uncooperative patients.

Follow a logical approach that can give you the maximum information quickly.

Most helpful -

Inspection- Note the findings as mentioned in the *general evaluation* and the *inspection* of the chest section

Auscultation- it can be done in a parent's lap or shoulder

Least helpful - Palpation and percussion.

PHYSICAL SIGNS IN PULMONARY DISEASES

Disease Process	*Mediastinal-deviation*	*Chest Movement*	*fremitus*	*Percuss.*	*Breath sounds*	*Adventitious Sounds*
Consolidation	No	Reduced over area	Increased	Dull	Bronchial or reduced	Rales
Bronchospasm	No	Hyperexpansion with limited motion	Normal or decreased	Hyper-resonant	Normal to decreased	Wheezes or Rales
Atelectasis	Shift towards lesion	Reduced over area	Decreased	Dull	Reduced	None or rales
Pneumothorax	Tension deviates trachea to the opposite side	Reduced over area	None	Resonant	None	None
Pleural effusion	Deviation to opposite side	Reduced over area	None	Dull	None	Friction rub (sometimes)
Interstitial Process	No	Reduced	Normal to increased	Normal	Normal	Rales

Normal Respiratory Rate:

Age	*Range of Normal (breath/min)*	Rapid
Newborn	30 -50	More than 60
Infancy	20 -30	More than 50
Toddler	20 - 30	More than 40
Children	15 - 20	More than 30

Signs of respiratory distress:

1. Tachypnoea
2. Tachycardia
3. Rescissions (Use of accessory muscles of respiration: subcostal, intercostal, supraclavicular and suprasternal)
4. Reduced air entry on auscultation
5. Pallor
6. Sweating
7. Pulsus paradoxus
8. Inability to speak
9. Cyanosis
10. Irritability
11. Symptoms and signs of CO_2 retention:
    1. Confusion, drowsiness and later coma
    2. Warm and sweaty hands (peripheral vasodilatation)
    3. Bounding pulse

4. Coarse flapping tremor of the outstretched hands
5. Papilledema (cerebral vasodilatation) in chronic CO2 retention.

N.B. 5-11 signs of severe respiratory distress

Types of respiratory diseases:

Obstructive respiratory diseases:

Neonates or young infants:	Older infants and children:
- Choanal atresia - Vocal cord paralysis - meconium aspiration - Laryngomalacia - Aspiration syndromes - Gastro-esophageal reflux - Tracheo-esophageal fistula - Palatopharyngeal incoordination	- Bronchial asthma - Foreign body - Laryngotracheobronchitis - Epiglottitis - Bacterial tracheitis - Adenotonsillar hypertrophy - Endobronchial tuberculosis - Vascular ring

Restrictive respiratory diseases:

Neonates and young infants	Older infants and children
- Pulmonary agenesis/hypoplasia - Hyaline membrane diseases - Diaphragmatic hernia - Congenital lobar emphysema - Severe eventration of the diaphragm - Asphyxiating thoracic dystrophy	- Pneumonia - Pneumothorax - Foreign body inhalation - Cystic Fibrosis - Pleural effusion - Neurological- - poliomyelitis - myasthenia gravis - botulism - muscular dystrophy - Skeletal- severe kyphoscoliosis - Obesity - Trauma - flail chest

Types of respiratory failure.

Definition - Respiratory failure is said to occur when:
PaO_2 < 60 mm Hg and/or
$PaCo_2$ > 50 mm Hg

Types:

Type I - PaO_2: low, $PaCo_2$: normal or low - *ventilation-perfusion mismatch*

Type II - $PaCo_2$: raised, PaO2: low - *ventilatory failure*

Mode of breathing in different diseases:

1. ***Obstructive:***
   - Mild: reduced rate, increased tidal volume
   - Severe: increased rate, retractions, anxiety, cyanosis
2. ***Restrictive:*** Reduced rate increased tidal volume
3. ***Kussmaul respiration:*** Increases rate, increased tidal volume, deep respiration, and metabolic acidosis i.e., Diabetes mellitus
4. ***Cheyne-Stokes respiration:*** Gradually increasing tidal volume followed by gradually decreasing tidal volume and apnea. Due to CNS injury, depressant drugs, uremia, prematurity.
5. ***Gasping respiration:*** Slow rate, variable tidal volume. due to hypoxia, shock, sepsis.

Blood gas analysis:

PH		7.37	-	7.45
	Acidosis	<=======		=======> alkalosis
HCO_3		21	-	28 mmol/L
	Acidosis	<=======	metabolic	=======> alkalosis
PCO_2		35	-	45 mm Hg
	Alkalosis	<=======	respiratory	=======> acidosis
PO_2		85	-	100 mm Hg
	Hypoxia	<=======		=======> hyperoxia

*Base excess:*0 +/- 3

O_2 *saturation:* 95 - 99%

Methods of blood gas analysis:

1. ***Arterial blood gas*** *(ABG):* Most useful for pulmonary function.
2. ***Arterialized capillary blood gas:*** Correlates well with ABG. Local vasodilatation is achieved by warming a finger, heel or an earlobe.
3. ***Pulse oxymeter:*** Continuously measures peripheral oxygen saturation. Correlates well with arterial blood gas
4. ***Transcutaneous electrodes:*** Continuously monitor PO_2 and PCO_2 if tissue perfusion is adequate.
5. ***Capillary blood gas (CBG):*** More useful in measuring the chronic acid-base disturbances.

CLINICAL ANATOMY OF THE LUNGS:

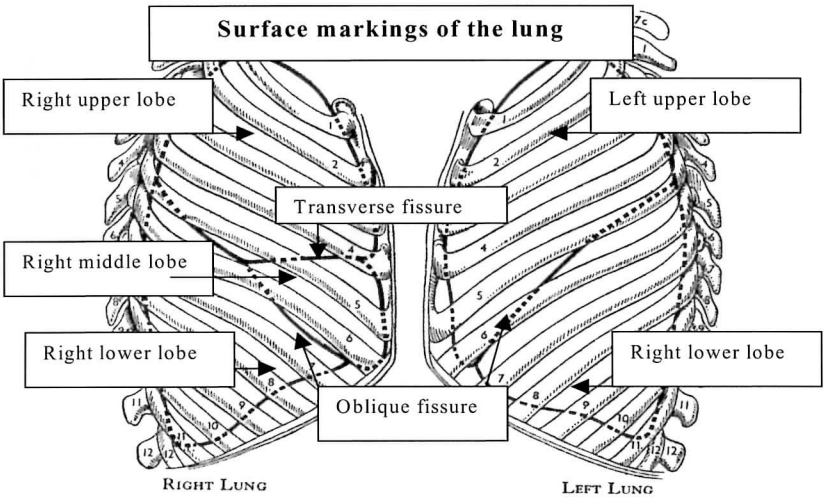
Division of the respiratory tract: (figure)

A) Upper respiratory tract - Includes nose, nasopharynx, larynx and extrathoracic trachea.

B) Lower respiratory tract - Includes intrathoracic trachea, bronchi and lungs.

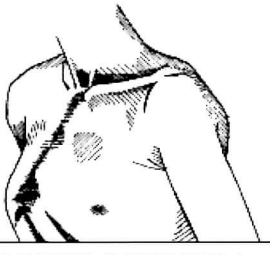
Right Lung - 3 lobes: - upper, middle, lower lobe.

Left Lung - 2 lobes: - lower and upper lobe including lingula (corresponding to the middle lobe of the right lungs)



CAUSES OF SOME IMPORTANT RESPIRATORY SIGNS:

Chest deformity: Pectus carinatum, Pectus excavatum and Harrison sulcus



PECTUS CARINATUM:
(Pigeon chest deformity)
Causes
- Asthma
- Other obstructive airway diseases
- Rickets
- Osteomalacia

PECTUS EXCAVATUM:
(Funnel-shaped deformity)
Causes
- Isolated congenital anomaly
- Chronic upper airway obstruction: (adenoid hypertrophy, laryngomalacia)

Causes of Harrison Sulcus:

- Chronic obstructive airway disorders- like asthma, cystic fibrosis, emphysema
- Bony weaknesses - rickets, osteogenesis imperfecta

Causes of deviation of mediastinum (trachea, apex)

a. *To the opposite side of the lesion:*
- Pneumothorax
- Pleural effusion
- Unilateral hyperinflation: foreign body, tumor causing bronchial obstruction
- Lobar emphysema: congenital or acquired

b. *To the same side of the lesion:*
- Collapse
- Fibrosis
- Hypoplasia

c. Heartbeat can be felt outside its usual position in case of the followings:
- Cardiac enlargement
- Dextrocardia - apex beat on the right side
- Scoliosis
- Diaphragmatic hernia

Causes of changes in the resonance on percussion:

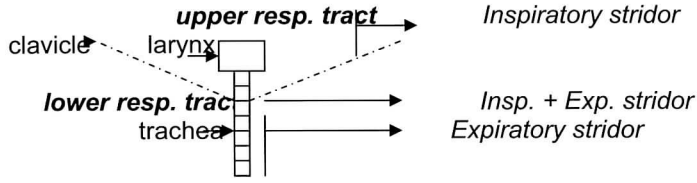
Decreased resonance:

1. Dullness on percussion:
   - a. Normal - Precordium
     - Hepatic dullness
   - b. Abnormal- consolidation
     - Fibrosis
     - Collapse
     - Pleural thickening
2. Stony dullness (absent resonance)
   - Pleural effusion

Increased resonance:
- Normal infant (thin chest wall)
- Pneumothorax
- Asthma
- Emphysema

Causes of bronchial breathing:
- Normal infant - on either side of the second thoracic spine at the back
- Normal children below the right clavicle
- Consolidation
- Large cavity
- At the air-fluid interface of small effusion (sometimes)
- Collapse

Origin of stridor:



Causes of acute stridor: *Remember ABCDEFG (not in order of importance)*

A. Angioneurotic edema, Anaphylaxis
B. Bacterial tracheitis
C. Croup (acute laryngotracheobronchitis)
D. Diphtheria
E. Epiglottitis (acute)
F. Foreign body inhalation
G. Rare:
    - Tetany
    - Trauma to larynx (burn, inhaled hot gas, mechanical trauma)
    - Peritonsillar abscess
    - Spasmodic croup

Causes of chronic stridor:

- Laryngomalacia
- Tracheomalacia
- Vocal cord paralysis
- Sub-glottic stenosis
- Laryngeal papillomatosis
- Vascular ring
- Laryngeal web
- Foreign body inhalation
- Sub-glottic or laryngeal hemangioma
- Laryngeal cleft
- Cyst - posterior to tongue or in the aryepiglottic fold

Causes of stridor in the first few days of life:

- Laryngomalacia
- Vocal cord paralysis
- Laryngeal web
- Congenital sub-glottic stenosis
- Vascular ring
- Hypocalcemia
- Macroglossia (Beckwith-Wiedemann syndrome)
- Choanal atresia

Differential diagnosis of wheezing:

- Asthma
- Bronchiolitis
- Aspiration syndromes-
    - Gastro-esophageal reflux
    - Tracheo-esophageal fistula (specially H-type)

  - Palatopharyngeal incoordination
  - Achalasia of esophagus
  - Pharyngeal pouch
  - Cystic Fibrosis, emphysema
- Bronchial obstruction - foreign body, tumor causing obstruction to a bronchus
- Congestive cardiac failure
- Angioedema

Not every tachypnea is due to pulmonary diseases, but it could be due to:

- Acidosis i.e., diabetes mellitus
- Fever
- Salicylism
- Extreme anxiety

Apnea:

Definition:
An apneic spell can be defined as cessation of respiration accompanied by bradycardia (heart rate<100 per minute) or cyanosis. Bradycardia and cyanosis are usually present after 20 seconds of apnea.

Types:

- *Obstructive*: absent airflow in the presence of inspiratory efforts, occurs less frequently.
- *Central*: inspiratory efforts and airflow cease simultaneously.
- *Mixed*: central pause is either preceded or followed by airway obstruction.

Causes of apnea:

- Apnea of prematurity
- Metabolic (hypoglycemia)
- Sepsis (meningitis – pneumonia - pertussis)
- Shock
- Drugs
- Severe anemia
- Hyaline membrane disease
- Persistent fetal circulation

BRONCHIAL ASTHMA

Working definition:
Bronchial asthma is a chronic inflammatory disorders of the airways in which many cells and cellular elements play a role. In susceptible individual, this inflammation causes recurrent episodes of wheezing, breathlessness, chest tightness, and coughing, particularly at night or in the early morning. These episodes are usually associated with widespread but variable airflow obstruction that is often reversible either spontaneously or with treatment. The inflammation also causes an associated increase in the existing bronchial hyper-responsiveness to a variety of stimuli.

Airflow limitation in asthma is recurrent and caused by a variety of changes in the airway. These include:

a. Acute bronchoconstriction: IgE and non-IgE dependant mechanisms.
b. Airway edema: due to increased microvascular permeability.
c. Chronic mucus plugs formation.
d. Airway remodeling interferes with response to treatment and causes persistent symptoms.

Key indicators for considering a diagnosis of asthma:

Wheezing: lack of wheezing and a normal chest examination do not exclude asthma.
History of any of the following:

- Cough, worse particularly at night
- Recurrent wheeze
- Recurrent difficulty in breathing
- Recurrent chest tightness
- Reversible airflow limitation and diurnal variation as measured by using a peak flow meter
- Symptoms occur or worsen in the presence of some precipitating factors (see later.

Differential diagnosis

- Foreign body in trachea or bronchus
- Vocal cord dysfunction
- Vascular rings or laryngeal webs
- Laryngotrachiomalacia, tracheal stenosis, or bronchostenosis
- Enlarged lymph nodes or tumor
- Viral bronchiolitis or obliterative bronchiolitis
- Cystic fibrosis
- Bronchopulmonary dysplasia
- Heart disease
- Aspiration from swallowing mechanism dysfunction or gastroesophageal reflux

Assessment of severity of chronic asthma

Symptoms	<2/Month	2-3/Month	1-2/Week	Continuous
Exacerbation/ nocturnal symptoms	<1 /Month	1 /Month	2-3 /Month	>4 /Month
PEF between attacks	> 80%	> 80%	60- 80%	< 60%
PEF Variability	<10%	10-20%	10-20%	>20%
Classification	Mild Intermittent Step I	Mild Persistent Step II	Moderate Persistent Step III	Severe Persistent Step IV

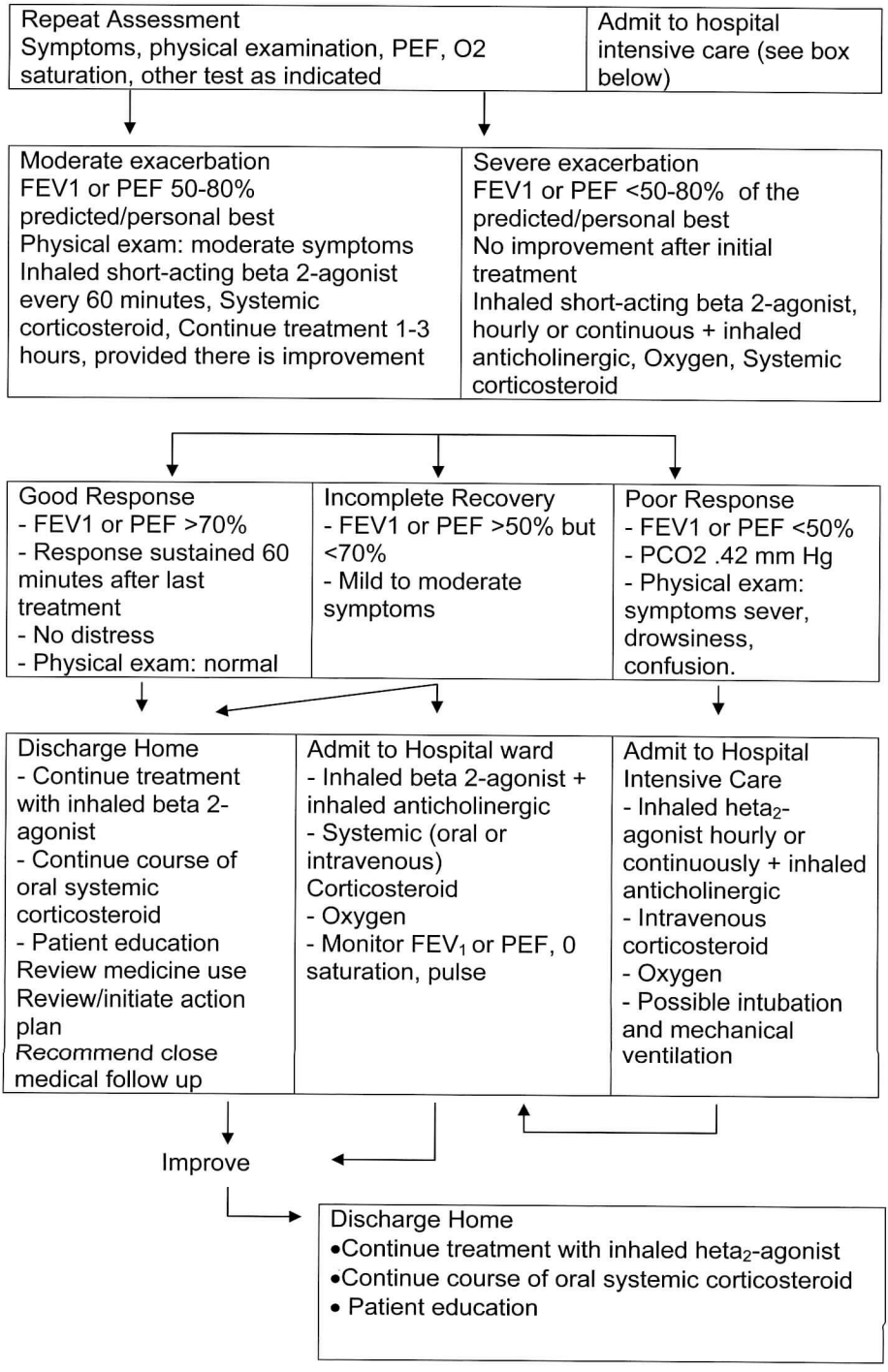
Alarming signs of severe acute attack of Bronchial Asthma

1. Cyanosis
2. HR > 120/min
3. Inability to speak in sentences
4. Silent chest
5. Use of accessory muscles
6. Subcutaneous emphysema
7. Significant differential reduction of breath sound suggesting mucus plug or pneumothorax
8. Agitation
9. Unable to lie flat
10. PEF after therapy less than 50%

Management of asthma exacerbations: emergency department and hospital-based care:

History, physical examination (respiratory rate, heart rate, use of accessory muscle, auscultation), PEF or FEV1, oxygen saturation, and other test as indicated.

FEV1 or PEF>50% - Inhaled beta 2-agonist by metered dose inhaler or nebulizer, up to three doses in first hour - Oxygen to achieve O2 saturation >90% - Oral systemic corticosteroid if no immediate response or if patient recently took oral systemic corticosteroid	FEV1 or PEF <50% (severe exacerbation - Inhaled high-dose beta 2-agonist and anticholinergic by nebulizer every 20 minutes or continuously for 1 hour - Oxygen to achieve O2 saturation >90% - Oral systemic corticosteroid	Impending or actual respiratory arrest - intubation and mechanical ventilation with 100% O2 - Nebulized beta 2-agonist and anticholinergic - Intravenous corticosteroid
↓	↓	↓


Repeat Assessment
Symptoms, physical examination, PEF, O2 saturation, other test as indicated
Admit to hospital intensive care (see box below)
Moderate exacerbation
FEV1 or PEF 50-80% predicted/personal best
Physical exam: moderate symptoms
Inhaled short-acting beta 2-agonist every 60 minutes, Systemic corticosteroid, Continue treatment 1-3 hours, provided there is improvement
Severe exacerbation
FEV1 or PEF <50-80% of the predicted/personal best
No improvement after initial treatment
Inhaled short-acting beta 2-agonist, hourly or continuous + inhaled anticholinergic, Oxygen, Systemic corticosteroid
Good Response
- FEV1 or PEF >70%
- Response sustained 60 minutes after last treatment
- No distress
- Physical exam: normal
Incomplete Recovery
- FEV1 or PEF >50% but <70%
- Mild to moderate symptoms
Poor Response
- FEV1 or PEF <50%
- PCO2 .42 mm Hg
- Physical exam: symptoms sever, drowsiness, confusion.
Discharge Home
- Continue treatment with inhaled beta 2-agonist
- Continue course of oral systemic corticosteroid
- Patient education
Review medicine use
Review/initiate action plan
Recommend close medical follow up
Admit to Hospital ward
- Inhaled beta 2-agonist + inhaled anticholinergic
- Systemic (oral or intravenous) Corticosteroid
- Oxygen
- Monitor FEV1 or PEF, 0 saturation, pulse
Admit to Hospital Intensive Care
- Inhaled heta2-agonist hourly or continuously + inhaled anticholinergic
- Intravenous corticosteroid
- Oxygen
- Possible intubation and mechanical ventilation
Improve
Discharge Home
•Continue treatment with inhaled heta2-agonist
•Continue course of oral systemic corticosteroid
• Patient education

Management of chronic asthma: Aim is to:

a. Minimize chronic symptoms.
b. Decrease frequency of exacerbation.
c. Minimize need for PRN B2 agonist medication.
d. Normalize activity.
e. PEF variability <15% (Almost normal PEF).
f. Minimal adverse reaction from medications.

Treatment of asthma should include:

a. *Treatment of the acute attack* Relievers: See treatment of acute attack.
b. *Prevention*
    1. Avoid triggering factors
        - Nonspecific irritant
            - Smoking
            - Wood smoke, organic compound (strong perfumes, household sprays, cooking oil)
        - Indoors allergens
            - House dust mites
                - Encasing mattresses and pillows
                - Washing blankets and bed linins once a week
                - Remove carpet
                - Avoid stuffed toys
            - Room air cleaning devices
            - Animal allergen: furred and feathered animal (CAT)
            - Cockroach allergen
            - Fungi: cleaning and avoid humidity
        - Outdoor allergens: pollens and fungi
    2. Pharmacotherapy prevention: Controller:
        - Sod. Cromoglycate
        - Steroid
        - Long acting beta 2 agonist
            - Solometrole
            - Formoterol
            - Leukotrines modifier
c. Produce an action plan
    Every patients should have an action plane: it is a plane for the patients and his family to be done in case of acute attack that include initial assessment of the acute attack and accordingly what the patient should do from the followings: tacking bronchodilators, taking steroid, contacting his doctor *or going to* the hospital.
d. Follow up and referral to a specialized center.

Summary of Asthma medication:

Daily prophylactic medic.	***Rescue med.***	
Non	SA-B2 Agonist	Step I
Beclomethasone /Intal	SA-B2 Agonist	Step II
Leukotrines modifier (Oral)?	SA-B2 Agonist	Step II
Budesonide	SA-B2 Agonist	Step II
Fluticasone or budesonide	SA-B2 Agonist	Step III/IV
Fluticasone or budesonide and LA-B2 Agonist √ √	SA-B2 Agonist	Step III/IV
Fluticasone or budesonide and Leukotrines modifier (Oral)	SA-B2 Agonist	Step III/IV
Systemic steroid should be given for severe or prolonged acute attack irrespective to step classification SA = short acting		

1. Start with high dose to get asthma under control then reduce the doses as required.
2. Using single prophylaxis medication is preferable than two medications.
3. Inhaled steroid well give better effect but compliance with oral Antileukotrines is higher.
4. Neither LA B2 Agonist nor Antileukotrines should be given without inhaled steroid in step 3 or 4 asthma.

Special forms of asthma

1. Cough variant asthma:
   - Chronic cough > 2 weeks
   - No apparent cause
   - CXR, spirometry: normal
   - R, empirical trial of bronchodilators and/or anti-inflammatory medication
2. Nocturnal asthma:
   - Chronic cough at night
   - Sign of poorly controlled asthma
   - R, exclude triggering factors, long acting bronchodilators, anti-inflammatory agent
3. Exercise induced asthma:
   - Exercise induces bronchospasm in 75-80% of asthmatics.
   - Do not discourage exercise
   - Give two puffs of sulbutamol, 10-20 minutes prior to exercise or two puffs of cromolyn
   - Warm up period of 10 minutes is necessary

   - Leukotrines antagonist might have good effect
4. Aspirin- induced asthma:
   - Avoid aspirin and other NSAIDs
   - Desensitization can be tried if necessary
   - Leukotrines modifier might have a role
   - Other medication should be avoided: paracetamol, nitrofurantoin, b-blockers, vinblastine, amphotricine B, cisapride

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- Current Pediatric Diagnosis & Treatment, Hathaway, Groothius, Hay, Paislay, 10th edition.
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GASTROINTESTINAL SYSTEM

HISTORY:

If you are dealing with a child with gastrointestinal problems, in addition to the points already mentioned in the chapter of general history you have to ask some specific questions. For example - vomiting, diarrhea, blood in the stool, constipation, abdominal pain, jaundice, weight loss, dietary history, change in the behavior, faecal soiling, any extra-intestinal manifestations (like joint swelling, mouth ulcers and skin rashes in inflammatory bowel disease).

If it is a chronic condition, ask for previous investigations, surgical procedures, if any, and indications and results of these. History of drug intake, response and complications to it. Ask any history of dietary manipulations. Know about the antenatal history like polyhydramnios and premature delivery (congenital chloride diarrhea). In the neonatal history ask about umbilical catheterization (portal hypertension), time of passage of meconium (delayed in cystic fibrosis, Hirschsprung's disease)

EXAMINATION:

General Comments:

- **Nutritional status:**
  - Obese, thin or normal, signs of muscle wasting (flat buttock, loose skin folds in the thigh and axillae due to loss of subcutaneous fat).
- **Hydration status**.
- **Hands**
  - Clubbing,
  - Koilonychia (spooning of the nails) in iron deficiency
  - Palmar erythema e.g. chronic liver diseases
  - Flapping tremor e.g. liver failure
  - Purpura e.g. chronic liver disease
- **Eyes**
  - Jaundice - look at the upper bulbar conjunctiva while the child is looking downwards
  - Pallor- look at the lower palpebral conjunctiva while the child is looking upwards
  - Periorbital edema e.g. nephrotic syndrome, nephritis, other causes of hypoproteinemia.
- **Face, neck and upper chest**
  - Spider naevi (in area which is drained by the superior vena cava)
- **Lips**
  - Pigmentation in Peutz Jeghar's syndrome.
  - Cheilitis (inflammation of the lips)
  - Angular stomatitis e.g. riboflavin deficiency
- **Teeth**
  - Number of teeth, missing teeth, caries discoloration (yellowish in tetracycline, chalk-white in fluorosis) - dental abscess.
- **Gum**
  - Gingivitis (inflamed easily bleeding gum) hypertrophy e.g. phenytoin therapy, poor dental hygiene, stippled blue line along the edge of the gum in lead poisoning.

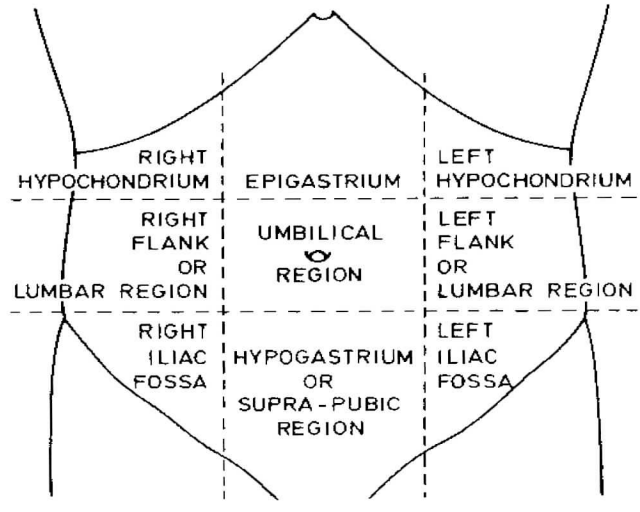
- **Tongue**
  - Ulcers
  - Color: pale and smooth in iron deficiency anemia
  - Strawberry tongue e.g. scarlet fever, Kawasaki disease
  - Geographic tongue: a benign condition.
- **Buccal mucosa**
  - Ulcers
  - Monilial thrush
  - Koplick's spot in measles
- **Palate, fauces, tonsils and pharynx:**
  - Cleft
  - Petechiae e.g. infectious mononucleosis
  - Ulcers
  - Vesicles e.g. herpangina, enanthem of varicella
  - Exudates over tonsils e.g. acute follicular tonsillitis, infectious mononucleosis

ABDOMINAL EXAMINATION:

To facilitate the description of the findings the abdomen is divided into nine regions by: -

1. Two horizontal planes-
   a) Subcostal
   b) Intertubercular
2. Two vertical planes which connect-
   a) Tips of the ninth ribs
   b) Femoral arteries just below the inguinal canal

The nine regions are:



INSPECTION:

- The child should lie flat and appropriately exposed from nipple to mid-thigh. Older children should be exposed up to the level of pubic symphysis and the genitalia should be examined after completion of the auscultation.
- Watch the environmental temperature.
- Observe from side and the foot end of the bed.
- Look for: -
    - Distention: e.g. fluid, flatus, feces and masses
    - Conduit: e.g. colostomy, iliostomy
    - Tubes: e.g. gastrostomy, nephrostomy, cystostomy, and peritoneal dialysis.
    - Restricted movement: e.g. peritonitis
    - Scars: e.g. previous peritoneal dialysis and surgery
    - Dilated veins: e.g. caput medusa in portal hypertension
    - Umbilicus: inverted (normal) or everted.
    - Obvious masses
    - Visible peristalsis
    - Hernias
    - -Genitalia.

PALPATION:

Please note: -

- Try to be at the level of the child's tummy
- Keep looking at the child's face to observe painful expressions
- Hands should be warm
- Ask the child where it hurts
- Palpate gently keeping hand flat and avoid poking with the finger tips

***Superficial palpation*:**

- Start from a point away from pain, if any.
- If there is no pain, start from the right iliac fossa and proceed clock-wise to end in the suprapubic area.
- Note: - Tenderness, Rigidity, and Masses

***Deep palpation*:** For

- Liver
- Spleen
- Kidneys
- Urinary bladder
- Any abnormal masses

Liver

- Start from the right iliac fossa; proceed towards the right costal margin. If liver is palpable, note the following:
- Measure the palpable liver in centimeter from right costal margin in the mid-clavicular line
- Nature of the border - whether sharp, irregular or rounded
- Consistency - soft, firm or hard
- Surface - smooth, nodular
- Tenderness
- Percussion - percusse over it and then percusse from the right second intercostal space in the mid-clavicular line to detect the upper border of the liver. Then measure the liver span in centimeter.

Spleen:

- Start from the right iliac fossa and proceed towards the left hypochondrium. If the spleen is not palpable roll the patient over to the right lateral position, splint the left lower rib cage with your left hand and palpate again while the patient is taking deep breaths. If the spleen is palpable:
- Feel for the notch
- Note the features as mentioned in case of liver

Kidneys:

Kidneys are:

- Better palpated bi-manually and they are also ballotable
- Put the left hand in the renal angle and right hand in the lumber area
- Push the left hand upward and the right hand downwards
- If palpable, look for ballotment
- Repeat the same on the other side
- Normally kidneys are not palpable except in the early infancy or in the very thin child where only the lower pole is palpable
- Kidneys are round and firm
- If palpable look for:
    - Size
    - Consistency
    - Surface
    - Tenderness
    - Percussion note - resonant.

***How to differentiate left kidney from spleen*:**

	Spleen	**Kidney**
1. To get above the swelling	-Not possible	-Possible
2. Notch	-May be felt	-Not felt
3. Percussion note	-Dull	-Resonant
4. Bimanually palpable and palatable	-No	-Yes
5. Movement with respiration	-Moves	-Does not move

Urinary bladder:

- A distended bladder is felt in the supra-pubic area
- It is globular arising from the pelvis
- Percussion note is dull
- Desire for micturition on handling.

Causes of distended bladder:

1. Normal child
2. Outflow tract obstruction: posterior urethral valve, stricture, stone
3. Neurological: spina bifida, neurogenic shock

***Palpate other areas*:** Note for-

- Tenderness
- Mass: -site
-Size
-Consistency
-Tenderness
-Movement with respiration
-Mobility, and
-Percussion note

PERCUSSION:

Look for- a) Ascites (*frequently asked in the examination*)
b) Percuss over any mass felt during palpation.

Ascites:

- **Fluid thrill**- *(for huge collection of fluid)*
  - Need one assistant, use either the patient's or examiner's hand, put the ulnar side of the hand in the middle of the abdomen
  - Put your left hand flat on the child's left lumber region
  - Now tap the opposite lumber region and feel the impulse by the flat of your left hand.
- **Shifting dullness**- *(for moderate collection of fluid)*
  - Start with the child in the supine position
  - Start from the umbilicus to-wards the left flank and note the point of dullness. Keep your finger at that point. Roll the child over the right side and wait for about 30 seconds.
  - Percuss over the same area and note the resonance. Now proceed towards the umbilicus till you get the dullness again i.e., the previous point of dullness is shifted from left to the right side i.e., shifting dullness.

- **Puddle test**
  - If the ascites in minimal it may be missed by the above methods, but may be elicited by putting the child in knee elbow position.
  - Percuss over the umbilicus and note the dullness, normally it is resonant.
  - Turn to supine position and percuss again note the resonance.

AUSCULTATION: For peristalsis and bruit

1. For peristalsis- over the right iliac fossa and one centimeter to the left and above the umbilicus
   a) Increased peristalsis- Early stage of intestinal obstruction
   b) Decreased or absent peristalsis- paralytic ileus or late stage of intestinal obstruction
2. Renal bruit- Auscultate on both sides over renal arteries, two centimeter lateral to the umbilicus, present in renal artery stenosis.

***Examination of genitalia*:** Examination procedure is discussed in detail in the chapter of endocrinology. However in short, inspect and palpate for -

- Ambiguous genitalia
- Scrotal pigmentation
- Micro or macro-penis
- Hypo or epi-spedius
- Urethral orifice
- Undescended testis
- Retractile testis
- Hydrocele
- Inguinal hernia
- Signs of puberty

***Examination of hernia*:** Hernia may be obvious during inspection, particularly in a crying child. The sites for hernia are inguinal, umbilical and epigastric areas. In a big child inguinal hernia is examined better in the standing position. Ask him to cough and look at the hernial orifices whether any swelling or pulsation appears. Try to get above the swelling. Examine for:

- Consistency
- Reducibility
- Tenderness
- Auscultation for peristalsis

Examine all the hernias in the same way.

Difference between inguinal hernia and hydrocele:

	Hernia	**Hydrocele**
Cough impulse	Transmitted	Not transmitted
Getting above the swelling	Not possible	Possible
Reducibility	May or may not be reduced	Not reducible
Transillumination	Negative	Positive

Rectal examination:

- This is to be done at the end
- Need not be routine in children
- Explain to the child and the parents
- Use lubricant
- Keep the child in the left lateral position with the knees flexed
- Indicated in:
    - Acute abdomen
    - Chronic constipation
    - Rectal bleeding
    - Suspected child abuse

Inspect for:

- Anal fissure: common in the 6 O-clock and 12 O-clock - positions
- Skin tags
- Fistula
- Fecal soiling
- Thread worms
- Signs of child abuse e.g., abrasions

Palpate: Gently introduce the finger and feel for:

- Anal tone: tight in anal stenosis and Hirschsprung's disease and absent in spina bifida
- Mass e.g., faecal mass, intussusception and appendicular mass
- Tenderness e.g., acute appendicitis
- Look at the finger-tip for blood stains.

Back: Don't forget to examine the back for:

- Scar
- Defect in the spine
- Swelling, tuft of hair or discoloration especially at the lower back

Examination of the lymph nodes and bones and joints:

If there is hepatomegaly and /or splenomegaly examine the *lymph nodes*:

- Cervical
- Axillary
- Inguinal, and also examine
- *Bones and joints* for any tenderness or swelling.

CAUSES OF ASCITIS:

1. Transudate-

a. Hypoproteinemia :
- Nephrotic syndrome
- Malnutrition
- Protein loosing enteropathy
- Hepatic failure

b. Hepatic causes:
- Cirrhosis
- Portal hypertension

c. Inferior vena caval obstruction:
- Hepatic vein thrombosis (Budd-Chiari syndrome)

d. Cardiac cause:
- Congestive cardiac failure
- Constrictive pericarditis

2. Exudate
- Peritonitis

3. Chylous
- Lymphatic obstruction or abnormalities:
  - Congenital- lymphangiectasia
  - Acquired - traumatic e.g., cardiac operations

ABNORMAL ABDOMINAL MASSES:

Right hypochondrium:
- Hepatic enlargement, Riedle's lobe of liver, gall-bladder mass

Left hypochondrium:
- Splenic enlargement, colonic mass

Right lumbar:
- Right renal mass
- Right suprarenal mass

Left lumbar:
- Left renal mass
- Left suprarenal mass

Right iliac fossa:
- Appendicular lump
- Crohn's disease
- Intestinal tuberculosis
- Ovarian cyst
- Ectopic or transplanted right kidney

Epigastric:
- Infantile hypertrophic pyloric stenosis (olive shaped mass elicited by a test meal)
- Choledochal cyst
- Pancreatic psuedocyst
- Bezoar.

Left iliac fossa:
- Faecal mass
- Sigmoid colon (in a thin child)
- Left ovarian cyst
- Left ectopic or transplanted kidney

Right sided sausage shaped mass with empty caecum - intussusception

HEPATOMEGALY:

In children, normal liver edge can be felt up to 2 cm below the right costal margin in the mid-clavicular line. The concept of normal liver size has been based on age and sex related clinical indices, such as

1. The degree of extension of liver edge below the costal margin.
2. The span of dullness to percussion.
3. The length of vertical axis of the liver which is estimated by imaging techniques.

Normal liver span:

Age: 1st. Week: 4.5 cm to 5 cm
12 years: Boys - 7 to 8 cm
Girls - 6 to 6.5 cm.

Causes of hepatomegaly:

- *Infection:*
  - Viral: e.g. Hepatitis A, B, C, D and E, EBV, congenital rubella, CMV
  - Bacterial: e.g. typhoid, brucellosis, tuberculosis, congenital syphilis,and neonatal infection from any bacterial etiology.
  - Protozoal: e.g. toxoplasmosis, malaria, schistosomiasia, and leishmaniasis.
- *Chronic hemolytic diseases*: e.g.,
  - Thalassemias
  - Sickle cell anemia
- *Collagen vascular diseases*: e.g.,
  - Juvenile chronic arthritis (systemic onset)
  - Systemic lupus erythematosus
  - Chronic active hepatitis, chronic persistent hepatitis
  - Inflammatory bowl diseases
- *Neoplastic disorders*: e.g.,
  - Leukemia
  - Lymphomas
  - Langerhans cell histiocytosis
  - Hepatoma
  - Neuroblastoma, nephroblstoma - secondary deposits
- *Metabolic and storage disorders*:
  - Carbohydrate metabolism e.g.,
    - Galactosemia
    - Hereditary fructose intolerance
    - Glycogen storage disorders
  - Amino acid metabolism e.g.,
    - Tyrosinosis
  - Urea-cycle disorders
  - Lipid disorders e.g.,
    - Gaucher's disease
    - Niemann-Pick disease
    - Gangliosidosis
    - Wolman's disease
  - Mucopolysaccharidosis
  - Alpha-1 antitrypsin deficiency
  - Peroxisomal disorder - e.g., Zellweger syndrome
- *Cardiac causes*: e.g.,
  - Congestive cardiac failure
  - Constrictive pericarditis
  - Inferior vena-caval obstruction
  - Hepatic vein thrombosis

- *Structural liver diseases*: e.g.,
  - Extrahepatic biliary atresia
  - Choledochal cyst
  - Intrahepatic biliary hypoplasia
  - Polycystic diseases
  - Congenital hepatic fibrosis

Causes of splenomegaly:

- *Infection*:
  - Viral: e.g., E. B. Virus, cytomegalovirus, and viral hepatitis
  - Bacterial: e.g., brucellosis, typhoid, syphilis, sub-acute infectious endocarditis, miliary tuberculosis, and septicemia
  - Protozoal: e.g., malaria, leishmaniasis, toxoplasmosis, and schistosomiasis
- *Hemolytic disorders*: e.g.,
  - Thalassemia
  - Sickle-cell anemia, hemoglobin SC disease
  - Hereditary spherocytosis
  - Autoimmune hemolytic anemia
- *Inflammatory and granulomatous conditions*: e.g.,
  - Systemic juvenile chronic arthritis
  - Systemic lupus erythematosus
  - Polyarteritis nodosa
  - Chronic granulomatous diseases
- *Neoplastic diseases*: e.g.,
  - Leukemias
  - Lymphomas
  - Langerhans cell histiocytosis
- *Storage diseases*: e.g.,
  - Gaucher's disease
  - Niemann-Pick disease
  - Mucopolysaccharidosis

Portal hypertension:

- (Normal portal venous pressure is 5 to 10 mm of Hg.
- Portal hypertension: if portal venous pressure is more than 20 mm of Hg).

Causes of portal hypertension:

- **Pre-hepatic**: Portal vein thrombosis (umbilical catheterization in neonatal period), stenosis, atresia and extrinsic pressure on the portal vein
- **Hepatic**: Various causes of cirrhosis, congenital hepatic fibrosis
- **Post hepatic**: Hepatic vein obstruction (Budd-Chiari syndrome), inferior vena-caval obstruction, cardiac- chronic congestive cardiac failure, constrictive pericarditis

Causes of hepatosplenomegaly:

As mentioned before in causes of hepatomegaly and splenomegaly. In general they can be grouped as follows.

- *Infection*: e.g. Viral, bacterial, protozoal
- *Hematological*: e.g. thalassemia
- *Malignancies*: e.g. leukemias, lymphomas

- *Storage diseases*: e.g. Gaucher's, Niemann-Pick, mucopolysaccharidosis
- *Congenital cirrhosis*
- *Collagen vascular diseases*
  N.B. *In Saudi Arabia most causes of hepatosplenomegaly are due to infection and hematological causes.*

Causes of bleeding per rectum:

- **Anal fissure** – (the commonest causes).
- Infection: Shigella, salmonella, campylobacter, E. histolytica.
- Inflammatory bowel diseases.
- Bleeding disorders: e.g., hemophilia.
- Inherited disorders: Familial polyposis coli, Peutz-Jeghar's syndrome, juvenile polyposis.
- Intussusception.
- Henoch-Schonlein purpura: secondary to vasculitis and intussusception.
- Meckel's diverticulum.
- Upper gastrointestinal bleeding.
- Pseudomembranous colitis.
- Necrotizing enterocolitis.

Differential Diagnosis of the Recurrent Abdominal Pain in Children

- *Associated with upper GI inflammation*
  - Gastroesophageal reflux disease (GERD)
  - Peptic ulcer
  - Helicobacter pylori gastritis
  - Nonsteroidal anti-inflammatory drug ulcer
  - Crohn's disease
  - Eosinophilic gastroenteritis
  - Parasitic infection (*Giardia*)
  - Henoch-Schönlein purpura

- *Motility disorders*
  - Intestinal pseudo-obstruction

- *Partial small bowel obstruction*

- *Extraintestinal disorders*
  - Chronic pancreatitis
  - Chronic hepatitis
  - Chronic cholecystitis
  - Ureteropelvic junction obstruction
  - Abdominal migraine
  - Psychiatric disorders

ETIOLOGY OF CHRONIC DIARRHEA IN CHILDREN

- **Infection and infestation**
  Bacterial
  Viral.
  Parasitic

- **Parenteral**
  Urinary tract infection

- **Postinfectious**
  Carbohydrate malabsorption
  Malnutrition
  Bacterial overgrowth

- **Dietary**
  Overfeeding
  Milk protein hypersensitivities:
    Soy protein hypersensitivity (other protein hypersensitivities)
  Malnutrition

- **Chronic nonspecific diarrhea**

- **Carbohydrate malabsorption**
  Congenital.
    Lactase deficiency
    Glucose-galactose malabsorption
    Sucrase-isomaltase deficiency
  Acquired
    Lactase deficiency
    Secondary disaccharidase deficiencies

- **Immune defects**
  Agammaglobulinemia
  Isolated IgA deficiency.
  Defective cellular immunity.
  Combined immunodeficiency.
  Acquired immunodeficiency syndrome (AIDS).
  Autoimmune enteropathy

- **Metabolic abnormalities**
  Familial chloride diarrhea.
  Sodium-hydrogen exchange defect.
  Abeta- and hypobetalipoproteinemia.
  Acrodermatitis enteropathica.

- **Endocrine**
  Hyperthyroidism
  Adrenal insufficiency.
  Diabetes mellitus

- **Hormone-secreting tumors**
  Ganglioneuroma

- **Small intestine**
  Celiac disease
  Intestinal lymphangiectasia
  Eosinophilic gastroenteropathy

- **Pancreas**
  Cystic fibrosis
  Shwachman syndrome

- **Anatomic lesions**
  Malrotation
  Partial small bowel obstruction
  Blind loop syndrome
  Short bowel syndrome
  Lymphoma
  Intestinal pseudo-obstruction syndrome

- **Inflammatory bowel disease**
  Ulcerative colitis
  Crohn's disease

- **Pseudomembranous enterocolitis**

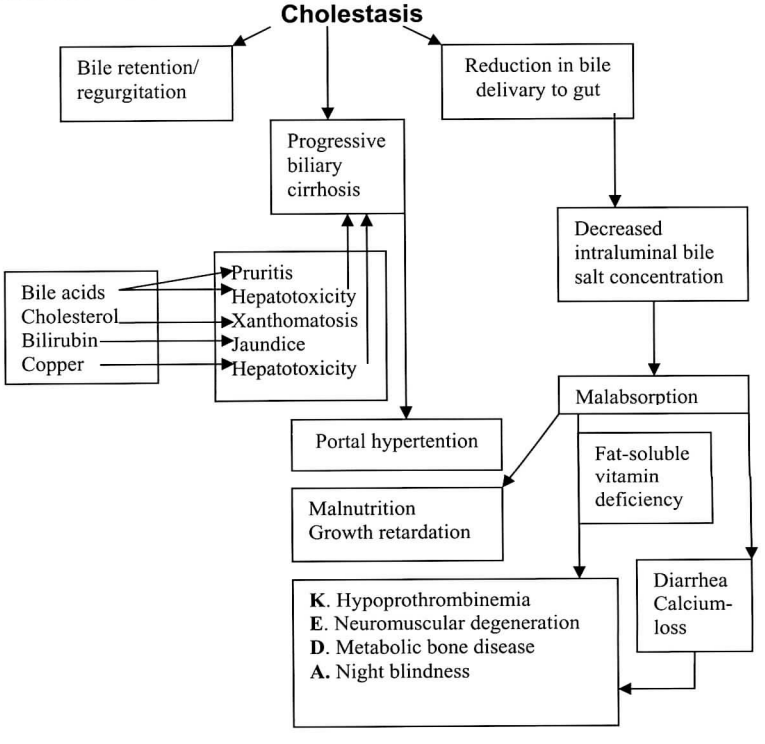
- **Antibiotic associated diarrhea**

- **Toxic diarrhea**

The Major Causes of Cholestasis in Infancy

- *Obstructive cholestasis:*
  - Biliary atresia
  - Congenital bile duct anomalies (choledochal cyst)
  - Cholelithiasis
  - Primary sclerosing cholangitis
  - Infectious cholangitis
- *Cholestasis with ductal paucity:*
  - Alagille's syndrome
  - Nonsyndromic ductal paucity
  - Ductopenic allograft rejection
- *Hepatocellular cholestasis:*
  - Hepatitis
  - α_1 - Antitrypsin deficiency
  - Inborn errors of bile acid synthesis
  - Drug-induced cholestasis
  - TPN-associated cholestasis
  - Progressive familial intrahepatic cholestasis

THE CONSEQUENCE OF CHOLESTASIS



DEHYDRATION:

Types of dehydration:

A. According to the severity:

	Percentage of body-weight lost deficit (per Kg)	**Volume**
Mild	3 -5 %	30 - 50 ml
Moderate	6 - 9 %	60 - 90 ml
Severe	=>10 %	100 ml or more

B. According to the serum sodium level:

a. Hypotonic: Serum sodium less than 130 mmol/L

b. Isotonic: Serum sodium from 130 - 150 mmol/L

c. Hypertonic: Serum sodium more than 150 mmol/L

***Clinical assessment of severity of dehydration*:**

The degree of dehydration can be estimated exactly by subtracting the present body weight from the previous body weight, if it is known. As this is not possible in most of the cases the following guidelines are used:

	Mild	**Moderate**	**Severe**
General appearance	Alert, thirsty	Thirsty, restless	Drowsy or lethargic
Pulse	Slightly rapid	Rapid	Rapid, weak, may be impalpable, poor capillary return
Blood pressure	Normal	Normal or low	Low or unrecordable
Tissue turgor	Normal	Absent	Absent
Anterior fontanelle	Normal	Sunken	Very sunken
Mucous membrane	Moist	Dry	Very dry
Tears	Present	Decreased	Absent
Eyes	Normal	Sunken	Deeply sunken
Urine output	Normal	Reduced and concentrated	Marked oliguria or anuria

Note:

In the hypernatremic status, classical signs of dehydration are less obvious, but CNS signs are prominent early. The child is usually irritable with high-pitched cry, doughy skin, full or even bulged anterior fontanelle and parched tongue.

ORAL REHYDRATION SOLUTION (ORS)

The composition of ORS as recommended by World Health Organization (WHO) is as follows:

	mmol/L		gm/L
Sodium	90	Sodium Chloride	3.5
Chloride	80	Sodium Bicarbonate	2.5
Bicarbonate	30	Potassium Chloride	1.5
Potassium	20	Glucose	20
Glucose	111		

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RENAL SYSTEM

A child with Renal disorder may present with obviously relevant symptoms such as hematuria or dysuria or part of systemic disease such as SLE or may remain a symptomatic for years while silent deterioration in renal function can occur.
The clinician must therefore be alert to the possibility of underlying renal disease, therefore diagnostic features of the history and examination worth emphasizing.

A) HISTORY

1) *Medical History*

- Clue to UTI
  - For example: fever, shivering, flank and back pain, cloudy offensive urine indicates upper UTI in older children.
  - Dysuria, frequent voiding and incontinence indicate lower UTI in older children.
  - Any previous similar attacks and investigations (for recurrent UTI).
  - History of chronic constipation
- Clue to Nephritis/ Nephrosis:
  - For example: coca-color or tea color urine, decrease urinary volume periorbital edema, respiratory distress, headache, hematuria, recent history of URTI or skin infection indicates post streptococcal glomerulonephritis.
  - Rash in the buttocks, lower limb, ± abdominal pain, ± arthralgia/arthritis, ± hematuria indicate H.S.P.
  - History of rash in the nose, cheek, arthritis/arthralgia, fever may indicate SLE.
  - Known to have congenital heart disease with fever, edema, coca color urine, decrease urine output, Headache may indicate subacute bacterial endocarditis and nephritis.
  - Hydrocephalic patient on AV-shunt with similar above clinical presentation may indicate shunt Nephritis.
  - History of generalized body edema, decrease urine output ± headache with urine dipsticks for protein >2+ indicate Nephrotic Syndrome.
  - Previous history of Hepatitis B or Malaria or Malignancy such as lymphoma generalized edema and proteinuria may indicate Nephrotic Syndrome 2^{o} to the above diseases.
- Clue to Chronic Renal Failure:
  May be silent and symptoms only developing late in its course such as:
  - Failure to thrive, vomiting, lethargy, behavioral changes indicate chronic uremia.
  - Short stature and delayed puberty.
  - Pallor (anemia) from chronic uremia.
  - Headache and convulsion (may indicate hypertension)
  - Polyuria indicates preterminal renal failure

- Decrease urine output, respiratory distress, edema in patient known to have chronic renal failure may indicate terminal renal failure or acute in top of chronic.
- History of dialysis or plan for transplantation

- Clue to Acute Renal Failure such as :
  - Any of the above previous symptoms in Nephritis and Nephrosis may lead to acute Renal Failure in a previously well child in addition to
  - History of recent cardiac surgery
  - Drugs such as antimicrobial, contrast media, history of anesthesia, cytotoxic drugs, cyclosporin.
- Clue to Renal Tubular Disorder
  - FTT/Growth retardation, polyuria, polydypsia, vomiting, constipation, fatigue, attacks of dehydration, delayed development/walking.
  - Use of drug like Furosemide, Vit.D, Dexamethasone, Cisplatin, Amphotericin B
- Others such as:
  - Gross Hematuria: check history of drugs such as cyclophosphamide, stones in urine, trauma or physical or sexual abuse, suprapubic aspiration or foley's catheter peritoneal dialysis, blood disease or had crash injury.
  - Poor urinary stream in male may indicate posterior urethral valve.
  - Urinary retention: check history of stones, tumor, trauma, sexual abuse.

2) Obstetric and neonatal history

Such as:

- Oligohydramnios, large placenta, perinatal asphyxia, premature delivery or low birth weight baby, TORCH infection especially syphilis during pregnancy (indicate congenital Nephrotic Syndrome)
- History of umbilical artery catheterization may lead to hypertension and /or chronic renal failure.
- Perinatal asphyxia might lead to gross hematuria, large kidneys and acute renal failure 2^{o} to renal vein thrombosis.
- Any antenatal ultrasound finding.

3) Family history

Such as:

- Deafness, eye abnormalities, chronic renal failure dialysis, transplantation, recurrent hematuria may indicate Alport's Syndrome.
- History of recurrent gross hematuria with no edema, convulsion or headache or renal failure may indicate idiopathic hypercalciuria
- History of stones in the family
- History of VUR
- Recurrent UTI
- Microscopic hematuria with no edema hypertension or renal failure may indicate benign familial hematuria.

B) EXAMINATION

1) General Examination

- Ill looking
- Level of consciousness
- State of hydration
- Edema puffy face or generalized in lower limb, sacral, scrotum in male labia majora in female and soft auricle
- Respiratory distress
- Check blood pressure in standing and sitting position if high for age check for signs of volume overload, heart exam and fundus.
- Inspect mucus membrane, conjunctiva and nail bed for signs of anemia.
- Skin and conjunctiva for jaundice.
- Nail for dystrophy point to nail patella syndrome
- Purpuric rash in lower limbs and buttocks for H.S.P.
- Rash in nose and cheek (SLE)
- Muddy complexion and signs of pruritis point to uremia in chronic renal failure.
- Caushengoid features, stria in lower limbs and lower abdomen, short stature and hirsutism points to side effect of steroid.
- Multiple cafe' aulet spots point to Neurofibromatosis for Renal artery stenosis and hypertension.
- Adenoma sebacuim and shagreen patches point to tuberous sclerosis for cystic kidney disease.

2) Head and Neck Examination

- Dysmorphic features such as deformed external ear may be associated with renal anomalies.
- Gingival hypertrophy points to cyclosporin toxicity
- Large tongue: Beckwith Wiedemann Syndrome (Renal Medullary Dysplasia)
- Separated, deformed teeth: Renal tubular disorder (Carbonic anhydrase deficiency Type II).

3) Cardiorespiratory System:

- Palpate femoral pulses may point to coarctation of the aorta with renal failure and hypertension
- Bruit over carotid and renal arteries for renal artery stenosis.
- Signs of cardiomegaly indicate volumes overload or long standing hypertension.
- Pericarditis with friction rub evidence of advance uremic state.
- Murmur point out to subacute Bacterial endocarditis lead to Nephritis.
- Rapid respiratory rate, recession, diffuse fine crepitations of lung basis indicates pulmonary edema.
- Kussmaul respiration: point to metabolic acidosis.

4) Abdomen: (please check abdominal examination in GIT)

Abdominal distention:

Check for ascites

- Large Kidney
- Retention of urine in bladder
- Absent abdominal wall musculature (Prune Belly Syndrome)

If abdominal distention not obvious, ask the child to lie in supine position and put his arms across the chest and raise his shoulder off the bed and see the upward movement of the umbilicus.

Renal Enlargement

Can be palpated in healthy neonate but not in older children

Unilateral

- Hydronephrosis 2^{ry} to PUJ obstruction
- Multi cystic kidney
- Renal vein thrombosis
- Malignancy
- Wilms Tumor

Bilateral

- Polycystic Kidneys
- Hydronephrosis 2^{ry} to posterior urethral valve, urethral structure
- Bilateral Renal tumor

Hepatosplenomegaly:

- SLE
- JRA
- Bacterial endocarditis
- Shunt Nephritis
- Hypersplenism with hepatic fibrosis in ARPKD
- Hepatomegaly $\rightarrow$ glycogen storage disease associated with glomerulus sclerosis.

Bladder: enlarged bladder should be examined by palpation and percussion.

If bladder detected ask the patient to pass urine and examine. In case of retention of urine, do rectal exam to exclude pelvic mass.

Genitalia

- Look for foreskin in male
- Position of urethral meatus for Hypospadias and epispadias.
- Position of Testes (undescended testes)
- Ambiguous genitalia

Anus: imperforated or abnormally positioned opening.

5) Musculoskeletal and CNS Examination

- Hypertonia & Hypotonia may lead to recurrent UTI and Nephrocalcinosis
- Muscle wasting in chronic Renal Failure

- Signs of Rickets for Renal Osteodystrophy or Renal tubular disease
- Meningomyelocele, Tuft of hair in the lumbosacral area may have recurrent UTI Neurogenic bladder and hydronephrosis
- Hemihypertrophy in Wilms tumor
- Facial nerve palsy → indicates severe hypertension and intracranial hemorrhage.
- Absent thumb and radius may indicate Fanconi anemia with Hypoplastic or Horseshoe Kidney.

6) Ophthalmologic Examination

- Papilledema (Hypertension)
- Cataract (side effect of steroid)
- Keratoconus in Alport Syndrome
- Retinitis pigmentosa (Juvenile Nephrolithiasis)
- Cystine crystal in the cornea (cystinosis) by slip-lamp examination
- Aniridia (Nephroblastoma)

C) URINE ANALYSIS:

Must be done as a part of routine evaluation of any child suspected to have renal disease.

Urine should be tested by:

1. Inspection for color and turbidity
2. Dipstick for * Blood
   * Protein
   * Glucose
   * PH
   * Nitrite

Microscopic examination for RBC's, RBC's casts, other casts, crystal. Specimen should be freshly voided sample maximum 30 minutes old, if delayed should be centrifuged and refrigerated at + 4° C to prevent bacterial over growth.

URINARY TRACT INFECTION (UTI)

(Please read also clinical approach to Renal System)

Diagnosis of UTI sometimes requires a high degree of suspicion because of the non-specific nature of the symptoms particularly in infants and young children. Proper diagnosis is important to prevent long term complications such as: **(1)** Renal scaring. **(2)** Hypertension. **(3)** Chronic pyelonephritis. **(4)** Chronic Renal Failure.

- ***Definition of UTI:***

  The growth of an abnormal number of bacterial colonies from the urine. From the above definition, Urine Culture may be the only confirmatory method for UTI patients provided that the specimen is obtained properly, transported to the laboratory quickly within 30 min from the collection and stored at a temperature ≤ 4°C until plated.

- ***Urine collection***

I. Preparation for collection

A) The perineum should be cleaned thoroughly with plain water without antiseptic to avoid false negative results.

B) In uncircumcised male the prepuce should be retracted before cleaning and collecting urine.

2. *Methods of collection:*

A) Supra pubic bladder aspiration: the best method in neonate and young children. Any number of colonies considered significant.
B) Intermittent "in and out" catheterization: especially in symptomatic patient required early treatment, $\geq 10^3$ colonies significant.
C) Mid Stream urine in toilet-trained patient, the first portion of the urine is to be discarded, the middle portion collected and the final portion discarded again. Positive culture if colonies $\geq 10^5$.
D) Bag urine collection: more likely to be contaminated, used mainly in non-toilet trained children. In this method a negative cultures rules out UTI considered to be significant if 2 out of 3 cultures positive with the same organism and colonies $\geq 10^5$.
- At least two independent samples must be collected. First morning specimen is the best.

- **Urine culture:**

To avoid false negative culture:
- Always checks:
  1) If patient on antibiotic during sample collection.
  2) If sample taken first in the morning.
  3) Way of preparation and collection.

To avoid false positive culture:
- Check
  1) Storage temperature
  2) Any vaginal infection in female child.
  3) Way of preparation

If culture grows proteus mirabilis, pseudomonas aeruginosa or Candida albicans it might indicate complicated cases.

- **Urine analysis:**
  1. Detection of pyuria by microscopic examination might suggest UTI in children but not diagnostic because UTI might occur without pyuria and pyuria can occur in many conditions such as:
     a) dehydration
     b) calculai
     c) trauma
     d) chemical irritation
     e) Renal tuberculoses.
  2. Detection of:
     a) Nitrite by dipstick might suggest presence of urea splitting organism like proteus, which convert nitrate to nitrite.
     b) Leukocyte esterase by dipstick, which release from rupture of leukocyte in urine might present but not diagnostic.

  Like in urine culture at least 2 early morning samples required for analysis.

- **Clinical evaluation of UTI patient**

Once diagnosis has been reliably made, it is necessary to determine:

1. The extent of the infection
2. Anatomy of UTI patient
3. Identify any condition that may predispose to recurrent infection.
4. If there is any associated complication such as chronic renal impairment, scaring, hypertension, renal failure.

For optimal further radiological investigation and treatment, it's easier to categorize UTI patients into 2 groups according to the Clinical presentation.

1) Asymptomatic
   - Discovered accidentally to have UTI especially in school age children.
   - At least 2 urine samples need to be cultured to confirm diagnosis.

2) Symptomatic
   A) Uncomplicated: lower urinary tract infection or cystitis.
   B) Complicated upper urinary tract infection or pyelonephritis.
   C) Associated with underlying disease such as:
      1. Obstructive uropathy
      2. Vesicoureteral reflux (VUR)
      3. Neurogenic bladder
      4. Voiding dysfunction
      5. Renal calculi
      6. Systemic disease such as diabetes mellitus, immunological defect
      7. Presence of an indwelling catheter or nephrostomy tube.

- **Radiological evaluation indicated in:**
  1. A first incident of asymptomatic bacteruria or UTI in a child younger than 5 years.
  2. First incident of asymptomatic bacteruria or UTI in any male child
  3. Recurrence of bacteruria or UTI in any female child
  4. Family history of UTI or urinary tract abnormalities
  5. Abnormal voiding pattern, poor growth or hypertension

HEMATURIA

- Please read also clinical approach to Renal Disease.
- Hematuria: it could be either gross (Macroscopic or Microscopic)
- Definition of microscopic hematuria: presence of ≥ 5 RBC/hpf on at least 2 properly performed centrifuged urine over a week.
- Mild 6-20 RBC/hpf, significant > 20/hpf
- The *first question* need to be answered if it is true hematuria or only colored urine due to:
  a) Drugs such as Rifampicin, Nitrofurantoin desferoxamine.
  b) Foods such as beet root and berries.
  c) Urate crystals
  d) Porphyrin
  e) Hemoglobinuria
  f) Myoglobinuria

To get answer in addition to history and clinical examination check urine for blood by (1) dipstick (2) microscopic examination:

- Positive hemostick and negative RBC'S by microscopic examination, it could be either due to hemoglobinuria or myoglobinuria.
- Positive RBC'S in microscopic examination means real hematuria

- The *second question* need to be answered, the origin of the hematuria if it is glomerular or non-glomerular.

Clues to localization of hematuria:

Glomerular

- Brown or tea colored (coca-colored) urine
- Red Blood cell lasts, cellular casts, tubular cells
- Proteinuria > 2+ by dipstick in the absence of gross hematuria
- Dysmorphic RBC'S by phase contrast microscopy
- Erythrocyte volume < 50 μm^2

Non-glomerular

- Red dark urine
- Blood clots
- No proteinuria or ≤ 2+ in the absence of gross hematuria
- Normal morphology of erythrocytes
- Erythrocyte volume > 50 μm^3

- The *third question* needs to be answered about the causes of hematuria.

Glomerular

- Acute post-streptococcal glomerulonephritis
- Hemolytic uremic syndrome, H.S.P.
- Focal segmented glomerulonephritis
- Membrano-proliferative glomerulonephritis
- Mesangio-proliferative glomerulonephritis
- Recurrent gross hematuria syndrome
  - IgA Nephropathy (Berger Disease)
  - Al port Syndrome
  - Benign familial hematuria

Non-glomerular:

- UTI bacterial (E. Coli), viral (adenovirus) protozoa (schistosomiasis)
- Obstructive uropathy e.g. PUJ obstruction, posterior urethral valve.
- Cystic kidney disease
- Renal vein thrombosis, hemangioma
- Hypercalciuria, calculi, nephrocalcinosis
- Renal tumors (Wilms tumor), leukemia
- Foreign body(in urethra or bladder)
- Drugs such as anticoagulants, antibacterials (such as Gentamicin, Ampicillin, Penicillin, Aspirin, Cyclophosphamide, indomethacin)
- Strenuous exercise
- Coagulopathy, sickle cell disease, trait.

- Factitious e.g. Munchausen syndrome by proxy.

PROTEINURIA

(Please read clinical evaluation of Renal Disease)
Normal daily urinary protein excretion in a febrile child is around 100 mg or 150 mg/m^2.
However there are age and sex differences in addition to diurnal variation.
There are several clinical methods to detect proteinuria.

- Urinary albustick detect only albumin and provide qualitative assessment of urinary protein excretion, which is graded as follows:

GRADE	Measurement (mg/dl)
- ve	No albumin
trace	10 to 20
1+	30
2+	100
3+	300
4+	1000 - 2000

- Quantitative measurement of urine protein excretion by collecting timed specimen over 12-24 hrs
  - Normal < 4 mg/m^2/hr
  - Abnormal 4-40 mg/m^2/hr
  - Nephrotic range: > 40 mg/m^2/hr
    Or >50 mg/kg/day
    Or > 1 gm/m^2/day
- Alternative measurement: Urinary protein in mg/dl to urinary creatinine (mg/dl) ratio (U.Pr/ U.Cr ratio) in random urine sample.
  - Normal { < 0.2 in older children; < 0.5 in the first few months of life }
  - 1 suspicious of Nephrotic Syndrome
  - 2.5 diagnostic of Nephrotic Syndrome

Clinical approach to proteinuria:

1. **Exclude false positive** test for protein by dipstick such as in:
   1. Gross hematuria
   2. Pyuria and bacteruria,
   3. Contamination of the urine with antiseptic such as chlorohexidine
   4. Drugs such as phenazopyridine
   5. If dip stick kept the urine too long
2. **Asymptomatic proteinuria**:
   When proteinuria discovered 2 additional first morning voided urine specimen should be examined, if only the initial sample is positive, child may have intermittent or transient proteinuria.

Or check urine by dipstick twice/day for week. First early morning sample as soon as the patient arise and the second just before the patient retires to bed these records may show:

a. Intermittent proteinuria
b. Orthostatic proteinuria: morning sample negative for protein and positive with varying concentration in the evening sample.
c. Persistent proteinuria: All samples contain protein in this case
    1. Do time urine collection over 24 hrs for protein or u Pr/ u Cr ratio to qualitative severity of proteinuria
    2. Examine several urine samples by microscopy for hematuria.

3. **Symptomatic proteinuria:**
    - Children with severe proteinuria, periorbital or extremity edema must be evaluated to rule out Nephrotic Syndrome.
    - Other evidence of Renal Diseases such as hematuria, edema, polyuria, oliguria, dysuria, colicky abdominal pain, family history of renal failure or deafness.
    - Clinical evidence of growth failure, hypertension, anemia, renal tenderness or enlargement, renal osteodystrophy with proteinuria and hematuria. Need further investigation according to suspected etiology.

Causes of proteinuria

I. Intermittent proteinuria

A) Such as fever, dehydration, exercise, cold exposure, congestive heart, failure, seizures, emotional stress, epinephrine therapy.
B) Orthostatic (postural) proteinuria

II. Persistent proteinuria

A) Glomerular diseases
    - Glomerulonephritis
    - Post infectious (post streptococcal, hepatitis B associated, chronic shunt infections and subacute bacterial endocarditis).
    - IgA Nephropathy, H.S.P, focal segmental glomerulosclerosis, membranoproliferative GN, SLE, sickle cell anemia.

B) Tubulointerstitial disease:
    - Reflux Nephropathy, tubulointerstitial Nephritis, Cystinosis, Lowe syndrome, drugs (analgesic) and heavy metals.
    - Ischemic tubular injury, renal hypoplasia/dysplasia.

RENAL TUBULAR ACIDOSIS (RTA)

(Please see also clinical approach to Renal System)
Normal infants and children generate 1-3 mEq/kg/day acid from dietary protein and metabolism, this acid must be excreted by the kidney to preserve acid base hemostasis by re-absorption of filtered bicarbonate in the proximal tubules and excrete acid as titratable acid and ammonium in the distal tubule. When there is defect in the re-absorption of bicarbonate or excretion of hydrogen ion or both, systemic metabolic acidosis will develop this clinical syndrome called Renal Tubular Acidosis (RTA)

Classification of RTA:

1. Proximal RTA (type 2): defect in tubular re-absorption of bicarbonate.
2. Distal RTA (type 1) L defect in tubular excretion of H^+ in distal tubules.
3. Hyperkalemic RTA (type 4): deficiency or tubular insensitivity to aldosterone.

These classifications are not standard for every patient; variations, subgroups and combined defects are well recognized.

Clinical approach to RTA

Clinical Presentation:

- Symptoms are non-specific and high index of suspicion is necessary.
- In addition to the clinical presentation mentioned in the clinical approach to renal disease, patient might present with growth retardation (universal), or a picture of life threatening acidosis.
- Nephrocalcinosis, calculi and rickets, might discovered by renal ultrasound especially in type 1 RTA.
- Might present with musculoskeletal complains such as Myalgia and Muscle weakness inability to walk due to hypokalemia.
- Family history of similar illness: deafness, stones, nephrocalcinosis.

Diagnostic approach:

1. If RTA suspected, confirm it by doing:
   - A) Blood gases, blood urea, creatinine and electrolytes, If there is metabolic acidosis and high chloride, make sure that the patient not on:
     1. Parenteral nutrition
     2. Drugs such as salicylic acid
     3. Having renal disease such as renal failure with azotemia and acidosis
     4. Gastroenteritis
     5. Bladder augmentation by bowel segment
     6. Diabetic ketoacidosis
   - B) Repeat again blood gases, blood urea, creatinine, s. electrolytes if there is still metabolic acidosis and high chloride.
   - C) Calculate the anion gap in the blood =$(Na + K) - (Cl + HCO_3)$ if normal (9-13 mmol/l) go to the next step.
     (Note: Anion gap high in the above cause s of metabolic acidosis).

2. Evaluate site of Renal Tubular lesion:
   A) Check urine pH by pH meter from freshly voided morning urine collected under mineral oil to prevent loss of CO_2.
   B) Calculate urine anion gap by measuring sodium, potassium and chloride in the urine uAG = Na + K – Chloride (uAG = indirect index of urinary ammonium excretion).
   C) Conclusion:
      1. If urine pH <5.5, urine anion gap negative (high urine ammonium) i.e. chloride > Na + K, K low or normal lesion most likely in proximal tubules.
      2. If urine pH > 5.5, urine anion gap positive (low urine ammonium) Na + K > Chloride. S. Potassium low/normal or high lesion in distal tubules.
      3. If urine pH <5.5, urine anion gap positive, high serum K RTA due to deficiency or resistance to Aldosterone.

CAUSES OF RENAL TUBULAR ACIDOSIS:

PROXIMAL	***DISTAL***	***(TYPE 4)***
I. *ISOLATED*	***I.*** *PRIMARY*	***I.*** *Hypoaldosteronism*
- primary (sporadic or familial)	- Sporadic or familial	▪ Diabetes
- carbonic anhydrase deficiency		▪ Interstitial nephritis
- carbonic anhydrase deficiency Type II	***II.*** *Hereditary*	▪ Nephrosclerosis ▪ Amyloidosis
	• Marfan syndrome	▪ SLE
II *Generalized:*	• Ehlers-Danlos	▪ Transplant rejection
• Primary (sporadic or familial)	• Sickle cell anemia	▪ Analgesic abuse
• Hereditary Cystinosis	***III.*** *Disorders of Calcium Metabolizing and Nephrocalcinosis*	▪ Non steroid anti-inflammatory drugs ▪ Spironolactone
• Hereditary fructose intolerance	o Idiopathic or familial	
• Lowe Syndrome	o Idiopathic hypercalcemia	***II.*** *Aldosterone-Resistance states.*
• Galactosemia	o Hyperparathyroidism	- Obstructive uropathy
• Tyrosinemia	o Hypernatremia	- Tubulointerstitial disease pseudo
• *Wilson Disease*	o Medullary sponge kidney	- Hypoaldosteronism (type I & III)
• Medullary cystic disease		
	IV. *Drugs & toxic*	
III. *Acquired*	- Amphotericin B	
oOutdated tetracycline	- Ameloride	
oHeavy metals	- Analgesic Abuse	
oProteinuria		
oRenal transplant rejection	***V.*** *OTHERS*	
oHyperparathyroidism	▪ UTI, SLE	
oVit. D deficiency rickets	▪ SCA, Renal transplant rejection	

ACUTE RENAL FAILURE (ARF)

Definition: Sudden decrease in renal function accompanied by the retention of nitrogenous wastes such as blood urea nitrogen (BUN) and creatinine and disturbance of water and electrolyte balance.

Classification of ARF based on clinical criteria of urine output:

1. Oliguric (classic): urine output below 1 ml/kg/hr in neonate and young children and below 200 ml/m^2/24 hrs in older children.
2. Non oliguric: where there is normal or excessive urinary output >2 ml/kg/hr in the presence of an acutely rising BUN or serum creatinine.

Classification of ARF based on etiological factors:

1. Pre renal azotemia: Transient disturbance of renal function caused by hypo perfusion of the kidney.
2. Intrinsic or parenchymal azotemia: Prolong and severe hypo perfusion, nephrotoxic renal injury, intrinsic renal disease.
3. Post renal ARF: Acute urinary tract obstruction

Causes of ARF

I. *Pre-renal*

- Hypovolaemia: such as gastroenteritis, diabetic acidosis, hypoproteinemic states, hemorrhage, third spacing (peritonitis, ileus, burns).
- Peripheral vasodilation: sepsis, antihypertensive medication.
- Impaired cardiac output: congestive heart failure, pericardial tamponade.
- Bilateral renal vessel occlusion: artery, vein
- Drugs: prostaglandin synthetase inhibitors, angiotensin converting enzyme inhibitors, cyclosporin, diuretics.
- Others: increased intra abdominal pressure (from ascites) hepato-renal syndrome.

II. *Renal* (renal parenchymal azotemia in children)

- Circulatory insufficiency
- Nephrotoxine: (antimicrobials, contrast media anesthetics, heavy metals, organic solvents, cytotoxic agents, non-steroidal anti-inflammatory drugs.
- Disease of the Kidney or Vessels:
    - Acute glomerulonephritis associated with lupus, post strep. glomerulonephritis, rapidly progressive glomerulonephritis, IgA nephropathy.
    - Bilateral acute pyelonephritis, HUS, cortical or medullary necrosis, vasculitis, poly arteritis, hypercalcemia, hyperphosphatemia, hyper uricaemia, acute disease in the presence of chronic renal disease.
    - Myoglobinuria, hemoglobinuria, tumor infiltrate, intra tubular obstruction (sulphonamides uric acid, methotrexate)
    - Iatrogenic Factor: Removal of a solitary kidney, renal arteriogram, cardiac pump oxygenation.
    - Drugs: such as cyclosporin, nonsteroidal anti inflammatory drugs, converting enzyme inhibitors, antimicrobials (aminoglycoside, cephaloridine, amphotricine-B, Cotrimoxazol), diuretics.

III. *Post Renal*

- Posterior urethral valve, blocked bladder catheter, neurogenic bladder, surgical accidents
- Calculi, ureterocele, tumors in solitary kidney

Clinical approach to acute renal failure:

In addition to information mentioned in clinical approach to Renal Disease the following points should be considered:

A) History:

Ask about history of diarrhea, vomiting, hemorrhage, trauma, burn, urinary stream, drugs, previous abdominal surgery, costo-vertebra angle pain (suggest obstruction), bloody diarrhea.

B) Examination:

- Check for the signs of dehydration, trauma any site of bleeding, surgical scars.
- Measure blood pressure: hypotension indicates loss of vascular volume.
- Check the pulse and heart rate:
    1. Tachycardia indicates circulatory insufficiency.
    2. Slow irregular pulse indicates hyperkalemia.
- Carbopedal spasm indicates hypocalcemia.

C) Investigations:

Related to clinical presentation, the following serum and urinary indices might help to differentiate between pre renal and renal azotemia in oliguric well hydrated patient.

	Prerenal	Renal
BUN: Creatinine	> 20	<20
FENa	<1%	>2%
RFI	<1%	>1%
UNa	<20 mEq/L	>40 mEq/L
Specific Gravity	>1.020	<1.010
Uosm	>500 mOsm/L	<350 mOsm/L
Uosm: Posm	>1.3	<1.3

$$\text{Fractional Excretion of Sodium (FENa)} = \frac{\text{UNa} \times \text{Pcr.}}{\text{Ucr.} \times \text{PNa}} \times 100$$

$$\text{Renal Failure Index (RFI)} = \frac{\text{UNa (mEq/L) Pcr. (mg/dl)}}{\text{Ucr. (mg/dl)}}$$

Cr. = Creatinine
U = Urinary
Osm = osmolality
P = plasma

D. Challenge test:

Practical diagnostic test to differentiate between renal and pre-renal failure and it has therapeutic effect.

- Give 0.9% normal saline or 5% albumin 10-20 ml/kg /dose IV over one hour and observe the response within 2-3 hrs. Urine output should be > *1-3* ml/kg /hr if no response repeats with loop diuretics such as furosemide 2-5

mg/kg/dose IV if no response label patient as acute renal failure and manage as follows:

c) Do general supportive measure
d) Restrict fluid to insensible loss and output
e) Treat metabolic disturbance such as metabolic acidosis, hypocalcemia, hyperphosphatemia, hyperkalemia
f) Treat hypertension

CHRONIC RENAL FAILURE

Definition: Irreversible loss of renal function with a resultant decrease in the glomerular filtration rate (GFR), GFR < 20 ml / min. / 1.73 m^2

Stages of renal failure: GFR (ml /min. /1.73 m^2 SA)

1. Impaired renal function	80-50	asymptomatic
2. Chronic Renal insufficiency	50-30	Metabolic abnormalities Impaired growth
3. Chronic Renal failure	30-10	Progressive renal failure
4. End stage Renal failure	<10	Renal replacement therapy required.

Calculation of GFR

1. GFR in 24 hrs urine collection

$$GFR = \frac{UV}{P} \times \frac{1.73}{SA} = \text{ml /min. / 1.73 } m^2$$

U = urinary creatinine concentration (mg/dl)
V = volume of urine/min = Total urine volume (ml) divided by length of collection interval (min)
P = Serum creatinine concentration (mg/dl)
SA (m2) = surface area
Note: 24 hrs = 1440 min

2. GFR in spot urine

- Full term infant: (ml/min/ 1.73m^2) = 1.1 x body length (cm)

- Children: ml/min/1.73 m^2) = $\frac{K \times \text{body length (cm)}}{\text{S. Cr. (Mg/dl)}}$

S. Cr. = Serum Creatinine
K = constant
K in low birth Wt infant < 1 y = 0.33
Term <1y = 0.45
Children (2- 12 yrs) = 0.55
Normal value of GFR varies with patient age.

Causes of chronic renal failure:

- Glomerulonephritis: focal segmented glomerulosclerosis other glomerulonephritis
- Pyelonephritis/ interstitial nephritis: Obstructive uropathy, vesicoureteric reflux

- Hereditary (familial nephropathy: Polycystic kidney disease, medullary cystic disease including nephronophthisis, hereditary nephritis, cystinosis, primary oxalosis, and congenital nephrotic syndrome.
- Congenital Hypoplasia/dysplasia: Aplasia/hypoplasia/dysplasia; Prune - belly syndrome
- Multi system disease: lupus erythematosus, HSP, HUS.
- Renal vascular disease
- Miscellaneous: kidney tumor, drash syndrome
- Unknown

Clinical presentation:

Wide variety of presentation

1. May be silent progress insidiously and symptoms developed late such as failure to thrive, short stature, lethargy, pallor, UTI, enuresis, hypertension, congestive cardiac failure, hematuria.
2. Develop symptoms early such as in steroid resistant nephrotic syndrome. Renal mass, obstructive uropathy, failure of resolution of acute renal failure such as HUS

Points in favor of Chronic Renal Failure.

- Anemia
- Growth retardation
- Renal osteodystrophy
- Small kidneys by ultra sound

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ENDOCRINOLOGY & DIABETES

GROWTH

Definitions:

1) **Normal growth**: growth parameters between the 3rd and 97th percentile (may be equivalent to 2 ½ standard deviation below and above the mean respectively if normally distributed curves are used).

2) **Length**: stature of a supine child (usually < 2 years).

3) **Height**: stature of a standing child (usually > 2 years).

4) **Short stature**: length or height below the 3rd percentile.

5) **Tall stature**: length or height above the 97th percentile.

6) **Underweight**: weight below the 3rd percentile.

7) **Marasmus**: weight less than 60% of the ideal weight (50th percentile).

8) **Kwashirkor** presence of edema in a child weighing less than 80% of the ideal weight (50th percentile).

9) **Overweight** between 110 – 120% of the expected median percentile of the reference population (see below).

10) **Obesity**: more than 120% of the expected median percentile of the reference population best expressed on the percentage of the body mass index (BMI) calculated as:
1. $\frac{\text{Wt.}}{\text{Ht.}^2}$ $\frac{50^{th}\ \%\ \text{wt.}}{50^{th}\ \%\ \text{ht.}}$ X 100

11) **Growth velocity**: annual linear growth rate.

12) **Mid-Parenteral Height (MPH)**: $\frac{\text{Father height + Mother height} \pm 12.5}{2}$ cm. 3.(+ if a male & - if a female)

SHORT STATURE

Definition: see above
Etiology:

1. Genetic
2. Constitutional
3. Chronic illness e.g. chronic renal insufficiency, sickle cell anemia, celiac disease, etc.
4. Small for gestational age

5. Syndromes e.g. Turner's syndrome
6. Endocrinopathy :
    - Hypothyroidism
    - Growth hormone deficiency
    - Hypoparathyroidism
7. Social deprivation

Approach:

History:

- Onset
- Previous measurements of both weight and height, calculate growth velocity: normal is (25/year for 1st year 12.5/year for 2nd year then minimum 4 – 5 cm / year till puberty).
- Evidence of chronic illness
- Evidence of endocrinopathy: e.g. Neonatal hypoglycemia, Constipation, etc.
- Birth weight and mode of delivery
- Family history: parent's heights, family diseases, social deprivation

Examination:

- Accuracy of measurements (do it yourself, 2 people are needed to measure length).
- Evidence of chronic illness e.g, joint deformity, dysmorphism, etc.
- Endocrinopathy: e.g, goiter, midline defects, etc.
- Correct plotting: Be familiar with growth charts and know how to use them.
- Assess proportionism by:
    1) Measuring upper to lower segment (1.7:1 in newborns and almost 1:1 in adults).
    2) Arm span: This is usually equal to the height ± 5 cm.
- Fudoscopy: for optic atrophy.
- Goiter.
- Teeth.
- ENT: look for midline defect.
- Systems: look for evidence of chronic illness.
- Assess puberty as per Tanner staging.

Investigations:

- Bone age: hand and wrist x-rays. Comparison to standards e.g. Grenlich & Pyle Atlas.
- Blood count: Regarding anemia
- Renal function: Regarding chronic renal insufficiency
- Urinalysis: Regarding chronic UTI.
- Stool for ova and parasites.
- Antigliadin and endomyseal antibodies: Regarding celiac diseases

- Skull X-rays (pituitary views) : Regarding suprasella calcifications in
- craniopharyngioma
- Blood gases: Regarding renal tubular acidosis
- TSH, FT_4 to rule out hypothyroidism
- Chromosomes in girls to rule out Turners syndrome
- Growth hormone studies:
  - Random sample : useless
  - Physiological stimulus: (usually 1 is enough)
    - Exercise
    - Sleep
  - Biochemical stimulus: (usually 2 are enough)
    - Glucagon
    - Clonidine
    - Insulin – hypoglycemia
    - L. dopa – propranolol
    - L. arginine
    - Others

Treatment:

This is directed towards the cause.
Gastroenterology assessment may be needed.
Indications of growth hormone use are limited. They are:

1. Growth hormone deficiency
2. Turner' s syndrome
3. Chronic renal insufficiency

OBESITY

Definition – see above.
Aetiology:

1) **Simple, non-pathological, nutritional obesity. This constitutes the most common cause.**

2) **Endocrinopathy:**
   - Hypothyroidism
   - Cushing syndrome / disease
   - Hypoparathyroidism
   - Pseudohypoparathyroidism
   - Hyperinsulinism
3) **Syndromes:**
   - Prader - Willi syndrome (PWS)
   - Laurance – Moon Beidle syndrome (LMBS)

Management:

- If associated with tall stature, the most likely cause is simple (non-pathological) but think about hyperinsulinism.

- If the child is not able to remain euglycemic without feeding for more than one or two hours and if so, think about hyperinsulinism.

- Obese infants: think about over feeding.

- Obese adolescents: think about diet / lifestyle.
    - 16 – 20% of school children are obese.
    - 85 – 90% of obese children are not pathological.

- Look for specific dysmorphology eg.
    - Polydactaly in LMBS
    - Spindle shaped fingers in PWS
    - Short 4th metacapals in Pseudohypoparathyroidism
    - Goiter in hypothyroidism

If no cause is found, counseling is directed on diet and activity.

TALL STATURE

Definition - see above.

Causes:

- Constitutional
- Genetic
- Simple obesity
- Precocious puberty
- Gigantism
- Hyperthyroidism
- Syndromes: e.g.: Beckwith Weidmann, Weaver, Stoto, Kallman, Klinefelter, Marfan, Homocystinuria

Management:

Bone age to rule out significant advancement.
Investigation and treatment are directed by the clinical impression.
Rarely androgens are indicated.

CALCIUM DISORDERS

A) Rickets: A very common problem

Stages:

1) Stage I: Hypocalcemia
- Presentation: convulsions or asymptomatic
- Phosphate, alkaline phosphatase: normal
- PTH, X-rays: normal.

2) Stage II: Normal calcium
- Clinical and radiological changes present.
- Phosphate: normal or low
- Alkaline phosphatase and PTH: elevated.

3) Stage III: Severe changes as seen in stage 2 in addition to: hypocalcemia.

Set-up:
- 1 year old breast fed with no solid food or sun exposure.
- Adolescents with low calcium and Vit. D intake with minimal sun exposure with too much cola's.

Presentation:

Convulsions

Rachetic changes: (wide joints, wide ant. fontanelle, frontal bossing, craniotabes, rachitic rosaries, bowing), sweating, fractures (rare), delayed teething, delayed development, short stature, bone pains)

Types:
- Nutritional: Most common Vit. D deficiency in diet and/or lack of sun exposure.
- Malabsorption e.g. Celiac disease.
- Liver disorders: It has to be severe enough to cause malabsorption of Vit. D (Vit. D needs bile salt for absorption).
- Lack of hydroxylation in the kidney (Vit. D dependent D rickets type I, VDDR I)
- X-linked hypophosphatemic rickets due to hereditary renal tubular phosphate loss (also called Vit. D resistant rickets).
- Rickets associated with anticonvulsant use.
- Congenital lack of hydroxylation at liver level.
- Lack of tissue response to Vit. D (Vit. D dependent rickets Type 2, VDDR II).
- Hypophosphatasia: rare.

Diagnosis:
- **X-rays of wrist**: fraying, wide joint space, cupping, osteopenia, healing lines (if partially treated)
- **Chemistry:**
  - Ca: low or N depending on stage.
  - Ph: N or low depending on stage.
  - Alk. Phosphatase: High except in hypophosphatasia.
  - PTH: elevated in stage 2 and 3.

- 25 (OH) Vit. D_3 low except in VDDR I and hypophosphatemic rickets.
- 1, 25 (OH) Vit. D_3: the last to go down. High in VDDR II.

Treatment:

- Vit. D supplements to breast fed infants (400 IU daily).
- Daily and enough sun exposure and improvement of dietary habits.
- Treatment of the nutritional type is to give 10 times the requirement of Vit. D that is 4000 IU daily for 4-6 weeks.
- Treatment of other types is by use of active forms of Vit. D_3 and calcium or phosphate depending on type.

B) Hypocalcemia

Definition:

Calcium level < 2 mmol/L
< 1.8 mmol in neonates and preterms

Causes:

1) Rickets (see above)
2) Hypoparathyroidism
   i) Congenital:
      - Isolated
      - Part of Di George syndrome
      - Part of Catch 22 syndrome
      - Middle East Syndrome
   ii) Acquired:
      - Auto immune
      - Post thyroidectomy
      - Post neck radiation
      - Infections

Symptoms:

- Convulsions
- Numbness
- Irritability

Signs:

- **Chovestech sign**: Tape on the zygomatic bone of the face and observe the twitch (This however, can be normal in 10% of population).
- **Troussaus sign:** Carpopedal spasm if the BP cuff is inflated above systolic (usually 160 mm Hg) for 3 minutes.
- Signs of the cause (see rickets and hypoparathyroidism).

Treatment: If rickets see above.

- If hypoparathyroidism: 1 α Vit. D_3 and calcium.

THYROID DISORDERS

A) Hypothyroidism:

1. **Congenital hypothyroidism:**
   - Etiology:
     - Sporadic : Aplasia / Ectopic,
     - Autosomal recessive: Dyshormonogenesis
   - Usually asymptomatic at birth, jaundice, course facial feature, large tongue, umbilical hernia, lethargy, hypotonia, goiter, etc.
   - It is usually diagnosed by the screening tests (TSH of the cord blood). Thyroid scan differentiates the aetiology (Fig. 5).
   - Treated by L-thyroxine replacement for life.
   - Always treat, even if you are in doubt or if you feel it is transient for the 1st 3 years of life, confirmation can then be done after holding replacement safely at or before this age for 4-6 weeks.

2. **Acquired hypothyroidism:**
   - Most common cause is Hashimoto's thyroiditis which is autoimmune.
   - It presents with: goiter, pallor, weight gain, short stature, constipation, excessive sleep, rough skin, etc.
   - Diagnosis is made by demonstration of elevated TSH, normal or depressed T_4, and thyroid antibodies.
   - Treatment is with replacement of L-thyroxine for life.
   - Look for associations like diabetes mellitus, hypoparathyroidism, Addison's disease…. (Polyglandular disease).

B) Goiter:

Causes:

- Hashimotos Hypothyroidism
- Dyshormogenesis congenital hypothyroidism.
- Colloid simple goiter in adolescents.
- Multi-nodular goiter
- Gravis disease: rare in children
- Thyroid cancer: papillary, follicular, mixed, aplastic, medullary.

C) Graves disease : not common in children. It is diagnosed by clinical features (goiter, exophthalmos, hyperactivity, tachycardia, sleep disturbance, diarrhea, tall stature, weight loss, ……), elevated thyroxin level, depressed TSH, presence of thyroid stimulating immunoglobulins. Thyroid scan shows high generalized uptake (may be indicated to rule out thyroid nodule).

D) Thyroid cancer:

Rare

Types: papillary: follicular, mixed, aplastic, medullary.

Treatment: total thyroidectomy with radioactive iodine ablation followed by suppression L thyroxine treatment.

AMBIGUOUS GENITALIA

Definition: If clinical decision about the child gender is difficult to make, the child has ambiguous genitalia. Definition may also include bilateral undescended testicles, hernias in females especially if a gonad is palpable.

Sex differentiation:

1) **Genetic differentiation**: i.e. Fertilization.

2) **Gonadal differentiation:**
   - XY: The testes determining factor (SRY) will influence the gonad to differentiate to be a testicle.
   - XX: The gonad differentiates to be an ovary.

3) **Phenotypic differentiation:**

   a) **Internal organs**: The testosterone produced locally by the testicle will influence the male organs to develop (Wolfian tubes, epidydymis and seminal vesicles). The Mullarian inhibitory factor (MIF) produced locally by the testicle will inhibit internal female organs to develop. Vice-versa if the gonad is an ovary there is no testosterone, so the male structures regress and as there is no MIF so the Mullarian structures develop (fallopian tube, uterus, upper part of vagina).

   b) **External organs:**
      i) Phallus like structure will progress to be a penis in presence of normal systemic testosterone (stimulated by maternal placental HCG in the 1st trimester and fetal gonadtropine in the 2nd and 3rd trimester).

      ii) Labioscrotal folds fusion will occur if this tissue is exposed to androgens in the 1st trimester. (This process will not happen if stimulation occurs after 12 weeks of gestation).

      iii) Testicular descend - many factors are involved in this process.

4) Psychological adjustment:

Gender is realized by the child at 18th months of age. This is then strengthened by the secondary sexual characteristics at puberty.

Causes:

A) XX infant:

- **Fetal:** Adrenal: eg. cysts, tumors (rare), congenital adrenal hyperplasia (CAH) due to 21 hydroxylase or 11 hydroxylase def. (Fig. 6)

- **Maternal:**
  - External exposure to androgen.
  - Adrenal : Maternal CAH, cysts, tumors
  - Ovarian cysts or tumors

B) XY infant:

- **Pituitary :**
  - Panhypopituitarism
  - Isolated gonadotropine deficiency
  - Kallman syndrome
- **Testes :**
  - Testes vanishing syndrome
  - Biochemical defect in the androgen pathway e.g. 5 α reductase deficiency.
- **Tissues:**
  - Partial tissue unresponsiveness.
  - Complete tissue unresponsiveness (Testicular feminizing syndrome)

C) Mixed:

- Mosaic Turners
- Gonadal dysgenesis
- True hermaphroditism

Recently new nomenclature and definitions have been adopted by the Lawsin Wilkins Pediatric Endocrine Society (LWPES) and the European Society for Pediatric Endocrinology (ESPE) (Pediatrics Vol. 118, No. 2 August 2006). The term disorder of sex development (DSD) is the term used for intersex problems so that 46XYDSD should replace terms like male pseudohermaphroditism. Underviralization, and Undermusculanization. Similarly 46XXDSD should replace term like female pseudohermaphroditism, overviralization and masculinization. Ovotesticular DSD should replace the *term true* hermaphroditism.

Management:

On-call to see an infant with ambiguity, make no guess and do the following:

1) Brief history:
   - Similar family history (FH)

- FH of precocious puberty, delayed puberty, anosmia (inability to smell), irregular cycles.
- Maternal exposure to androgens.

2) Careful Examination:
   - General examination for major defects especially midline defects.
   - Local Exam:
     Describe what you see as follows:
     a) Phallus like structure: not penis or clitoris.
     b) Labioscrotal folds: not labia or scrotum.
     c) Gonad not ovary or testes.
     d) Inspect anus and examine hips.

3) Talk to the family and tell them that you can not decide and you need a few investigations to know and advice them not to give a name.
4) If you are not in a tertiary centre, transfer the baby to one.
5) If the child presentation is late and has a name already, continue as is till you know.
6) Ask for chromosomes (karyotype).
7) Monitor electrolytes (salt loss crises usually happen in the second week of life).
8) Ask for ultrasound and genitogram.
9) Ask for 17 (OH) progesterone, 11 Deoxycortisol, DHEA-S, Androstenedione, ACTH.
10) If XY chromosomes, you may need to stimulate with HCG and see the biochemical and the biological response. You may also need to stimulate with testosterone and see the biological response.
11) Specific treatment depends on the cause, eg., if CAH due to 21 hydroxylase def., replace with hydrocortisone, Florinef and sodium chloride orally (11 hydroxylase def: no need for Florinef or NaCl).
12) Treat shock and electrolyte imbalance accordingly.
13) Plan corrective surgery with the pediatric surgeon.
14) All XX infants will be raised as female.
15) Most XY infants are raised as males except if genitalia do not show biological response to androgens (See 10) like in testicular feminizing syndrome.
16) There is enough medial and Islamic support of the above recommendations.

HYPOGLYCEMIA

Definition: Generally < 2.2 mol/L (40 mg/dl) with symptoms which are relieved by glucose intake.

Aetiology:
- Ketotic hypoglycemia
- Endocrine:
    - Hyperinsulinemia
    - Hypopituitarism
    - Growth hormone deficiency
    - ACTH and/or cortisol deficiency (Adrenal insufficiency, CAH, etc.)
- Metabolic disorders
- Liver disorders
- Drug induced

History / Exam:
- Pattern of feeding.
- Symptoms and signs of hypoglycemia
    - Adrenergic: tremors, sweating, tachycardia, etc…)
    - Neuroglucopenia: headache, confusion, etc….)
- Dysmorphism: e.g. Macrocephaly, umbilical hernia, midline defects.
- Hyperpigmentation: e.g. in primary adrenal insufficiency

Investigations:
The critical sample is the sample to be taken during the presentation of the child with hypoglycemia: glucose, insulin, growth hormone, cortisol, ACTH, FT_4, lactate, pyruvate, hydroxybuterate, gases.

Treatment: This is directed towards the causes:
- If Hyperinsulinemia: Diazoxide, Nifedopine, somatostatin, pancriatectomy.
- If Hypopituitarism: Hormone replacement (growth hormone, etc…)
- If Ketotic hypoglycemia: frequent feeding and reassurance that prognosis is excellent.

PUBERTY

Definitions:

- **Normal puberty:** Development of secondary sexual characteristics at 8 to 13 years of age for girls and 9 to 14 years of age for boys.
- **Precocious puberty:** Development of puberty before the age of 8 years in girls and 9 years in boys.
- **Delayed puberty**: Lack of development of normal puberty by the age of 13 years in girls and 14 years in boys or 5 years lapse between onset and completion.
- **Thelarche:** Isolated breast development in girls. Isolated benign premature thelarche is the most common cause if this occurs before the age of 8 associated with no other puberty signs and with normal bone age.
- **Adrenarche:** Isolated sexual hair development. Isolated benign premature andrenarche associated with normal bone age is also a known phenomenon.
- **Menarche:** Onset of menstruation. It normally coincides with Tanner stage 4 puberty.
- **Spermarche**: Onset of sperm production.
- **Primary Amenorrhoea:** Delayed menarche beyond the age of 16 years.
- **Gynecomastia:** Breast development in boys, it is physiological at birth and at puberty.

Staging of puberty (Tanner staging)

Stage	Breasts	Male genitalia	Pubic hair for both girls and boys
1	No breast tissue	Testes < 2 ml volume.	None
2	Breast budding	Enlarging testicle >2ml reddening of scrotum.	A few hair along labia or penis
3	Areola and breast enlargement	Testes 6-10 ml in volume.	Curling of hair
4	Double angle, nipple and areola, and areola and breast.	Testes 10 – 15 ml in volume.	Hair over supra-pubic area.
5	Mature adult breast with loss of areola and breast angle.	Testes 15 – 25 ml.	Hair on inner thighs in addition to above.

PRECOCIOUS PUBERTY

Causes:

A) True (also called central)

- Idiopathic 50% in boys and 90% in girls.
- CNS insult : Trauma
- Tumors (eg. Hamartoma)
- Primary hypothyroidism
- Pseudo-precocious puberty

B) Pseudo-precocious (also called peripheral)

- Exogenious estrogens or androgens.
- Ovaries – cysts or tumors
- Adrenal hyperplasia, cysts, tumors
- Testes, tumor
- Liver tumors
- Mc Cunne Albrights syndrome
- Familial precocious puberty

Management:

History & Examination:

Geared into the aetiology. Points of interest: onset, is it normal or precocious? Small testicles favors pseudo-precocity, history of exposure to androgens or estrogens, review all systems e.g. headache or seizures which hints towards central aetiology, hepatomegaly, hyperpigmentation, skin hyperpigmentation eg. Café eu lait spots in Mc Cunne Albrights syndrome.

Investigation:

- Bone age : advanced
- FSH, LH, E_2, Testosterone
- LHRH stimulation test: if flat is normal prepubertal or pseudo - precocity but if positive in a <8 year old girl or 9 year old boy, it is central in type.
- **May need:**
  - Ultrasound of ovaries / adrenals / testicles.
  - 17 (OH) progesterone, compound(s), ACTH, electrolytes based on the suspected aetiological.
  - CT / MRI of the brain.

Treatment:

- **In pseudo-precocity**: treatment is directed towards the cause.
- **In true precocity:** also treatment is directed towards the cause in addition to LHRH analogue. For idiopathic types LHRH analogue is the drug of choice.

DIABETES MELLITUS

1) **Classification:**

a) Type I a. Immune mediated diabetes (IMD)
b. Idiopathic DM

b) Type 2 – obesity is a factor.

c) Gestational DM

d) Other specific types: e.g. genetic, endocrine related, etc…)

2) **Aetiology of Type I:** autoimmunity, genetic susceptible, syndromatic, infections, etc.

3) **Presentation:** polyurea, polydipsia, weight loss, abdominal pain, nocturnal enuresis, fever.

4) **Differential diagnosis:**

Polyurea, Polydipsia	Hyperglycemia
DM	DM
DI	Stress induced hyperglycemia
Chronic renal failure	
Hypokalemia	
Hypercalcemia	
Psychogenic	

5) **Management:**

Establish whether it is hyperglycemia only, hyperglycemia and ketonuria or both with acidemia (DKA)

A) Acute Diabetic Ketoacidosis.

1. Laboratory Investigations:

Initial: urinalysis, blood glucose, electrolytes, blood gases… others as indicated, blood gases and glucose alternating with electrolytes and glucose Q2 hr.

2. Fluids:

a) **If in shock (BP decreased):** Resuscitate like any other child with any fluid (preferably normal saline [NS]) at 10 – 20 ml/kg as quickly as possible. Consider plasma and/or albumin and possibly inotropic support (rarely needed). ICU care is usually needed. Once this is corrected proceed with rehydration as per the degree of dehydration and the electrolyte imbalance. (See below).

b) **If not in shock (normal BP):** Rehydrate the patient with 0.9 NS or 0.45 NS. As per the degree of dehydration and the electrolyte imbalance (beware of the polyurea, which may need 50% of urine output to be replaced);

- Degree of dehydration is 3, 6 and 9 percent for mild, moderate, and severe for children over 2 years of age and 5, 10 and 15 for children younger than 2 years of age.

- If the child is comatosed, offer the usual management of coma (airway, ventilation, IV line, N/G tube….etc).

- Keep the child NPO till pH is >7.3 and bicarbonate is >15mmol/L.

3. Electrolytes:

a) **Sodium:** 0.9 or 0.45 normal saline, continue with D5W 0.45 (preferred) or D5W 0.2 NS once the blood glucose is less than 250 mg/dl (children with DKA are total body sodium completed).

b) **Potassium:** Commence potassium chloride (or acetate or phosphate) as soon as possible (child voiding or serum potassium is normal). 4 - 6 meq. / Kg / 24 hours is needed. Rate should not

generally exceed 0.5 meq./kg/hr. Monitoring is needed.

c) **Bicarbonate:** This is rarely needed, fluid and insulin in most cases is enough to correct acidosis. Use if pH <7.1. Calculate deficit; Deficit x (wt. x 0.3 and give half of the deficit. (Usually acidosis is corrected before finishing this infusion so it does not have to be completed).

4. Insulin:

- IV 0.1 unit / kg regular insulin bolus, if continuous IV insulin cannot be prepared immediately.

- Infuse insulin at 0.1unit/kg/hr.
(Usually 50 units of insulin in 500 cc of NS and run at the same figure of the child's weight. This gives 0.1u/kg/hr.)

- Continue insulin dosage at that rates till acidosis is corrected (pH > 7.3 and / or bicarbonate > 15 mmol/L).

- If you run into hypoglycemia, insulin can be hold for 10 minutes and restarted at slightly lower but still supraphysiological dose, i.g. 0.05 units/kg/hr.

- Can decrease gradually to reach 0.02 unit/kg/hr. (physiological requirement) and leave till you are ready to switch to SC insulin (usually at breakfast or supper time)

- Once SC insulin is commenced at 0.5 unit/kg/day divided into 2/3 in the morning and 1/3 in the evening (almost 2:1 ratio of NPH to regular), IV insulin is discontinued ½ hr. After S/C insulin is administered.

5. Education

- Tell the diagnosis.
- Tell improvement will follow soon.
- Deal with patients psychology (Denial / Anger / Depression / Adaptation)
- Please refer to the ESPE/LWPEs consensus statement on diabetic ketoacidosis in children and adolescent (Dunger etal. Arch Di. Children; 89:188-194)

B. Diabetic Ketosis (with no acidosis):

If there is no acidosis and the child is not vomiting, ketosis can be managed by oral hydration and extra subcutaneous insulin according to the following sliding scale.

Blood Glucose (mmol/l)	Urine ketone	Regular insulin to be given (units per kg)
> 16	+++	1/4
> 16	++	1/6
> 16	+	1/8
> 20	0	1/8
< 16	+ / 0	None

This sliding scale is subjected to the circumstances at the time of assessment.

C. Teaching should include:

- Background information about physiology, absorption and metabolism of food, pathology in DM.
- Insulin: types, infections, etc.
- Testing: blood glucose home monitoring, urine for ketones.
- Hypoglycemia: Symptoms, measures to be taken, Oral sugar continuing fluid and if unconscious or seizing to utilize glucagon 30 μg/kg/dose which should be kept at home.
- Ilness management: Illnesses affect DM by two ways:

a) Lack of appetite, so frequent monitoring and sugar containing oral fluids to avoid hypoglycemia. Insulin may be decreased but never stopped.

b) Hyperglycemia and ketosis and if so supplement with extra regular insulin 0.1 unit/kg/dose every 4-6 hours.

D. Diet - A dietician is needed for this, give 1000 kcal for the 1st year then 100 kcal for every additional year and give it as 3 meals and 3 snacks.

Outpatient Care:

- Visiting every 3 – 4 months
- HbA1C every 3 – 4 months
- Check clinically and biochemically for the associated condition eg. hypothyroidism once every 1 – 2 years.
- Provide free axis to service.
- Continuous teaching.
- Group activities.

Complications:

- Retinopathy
- Neuropathy
- Nephropathy
- Failure of growth
- Others

DIABETES INSIPIDUS (DI)

Symptoms / signs: polyurea, polydepsia, dehydration, weight loss, etc.

Causes:

- **Congenital:**
  - Isolated
  - Panhypopituitarism
  - Nephrogenic
- **Acquired:**
  - Infections
  - Langerhans disease
  - Tumours : craniopharyngioma
  - Post surgery
  - Radiation
  - Trauma

Differential diagnosis of polyurea:

- DM
- Chronic renal failure
- Hypercalcimea
- Hypokalemia
- Psychogenic

Diagnosis: Water deprivation test.

Treatment:

- DDAVP
- Thiazide for nephrogenic type
- Chlorpropamide
- Cliofibrate

ADRENAL INSUFFICIENCY

Cause:

Primary:

- Autoimmune (Addisons)
- Infections Tb, HIV
- CAH
- Congenital adrenal hypoplasia
- Adrenoleukodystrophy

Secondary / Tertiary:

- Pituitary, hypothalamus

Presentation:

- Weight loss, fatigue, hyperpigmentation, vomiting, diarrhea, dehydration, etc.

Investigation:

- ACTH
- Cortisol
- ACTH stimulation test
- Search for the cause (Mantoux test, HIV screening, etc.)

Treatment: - Cortisol replacement

CUSHING DISEASE / SYNDROME

Presentation: - obesity, hypertension

Investigation:

- PM cortisol, ACTH.
- Dexamethasone suppression test, ultrasound, CT or MRI.

Treatment : - Surgery

Figure 5: Algorithm for TSH interpretation.

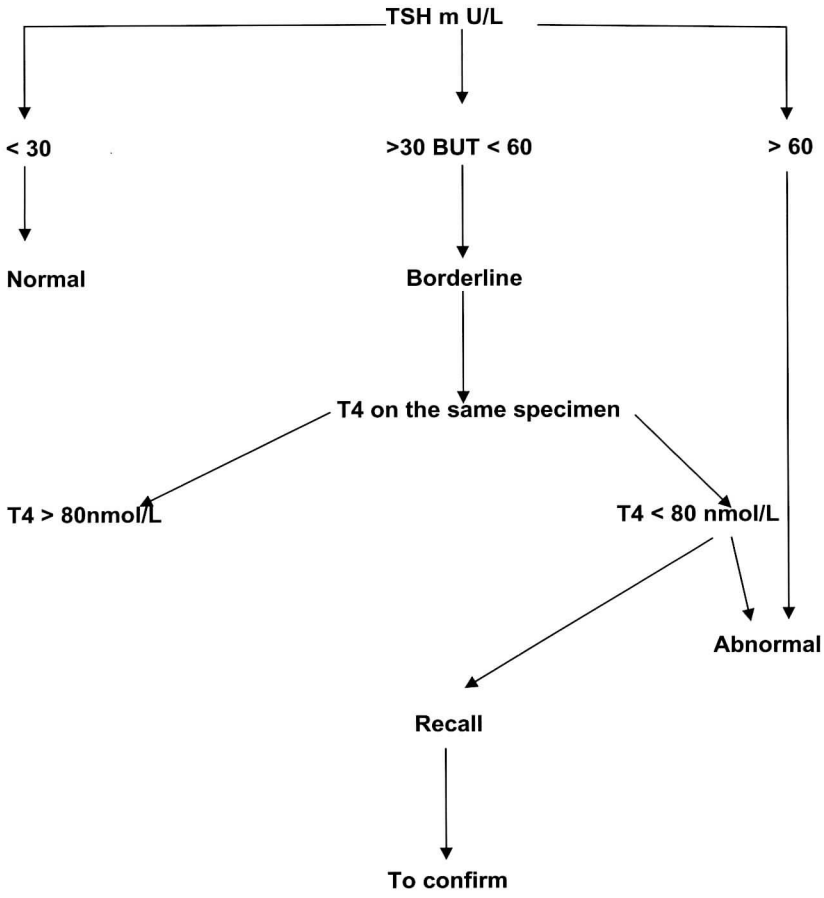
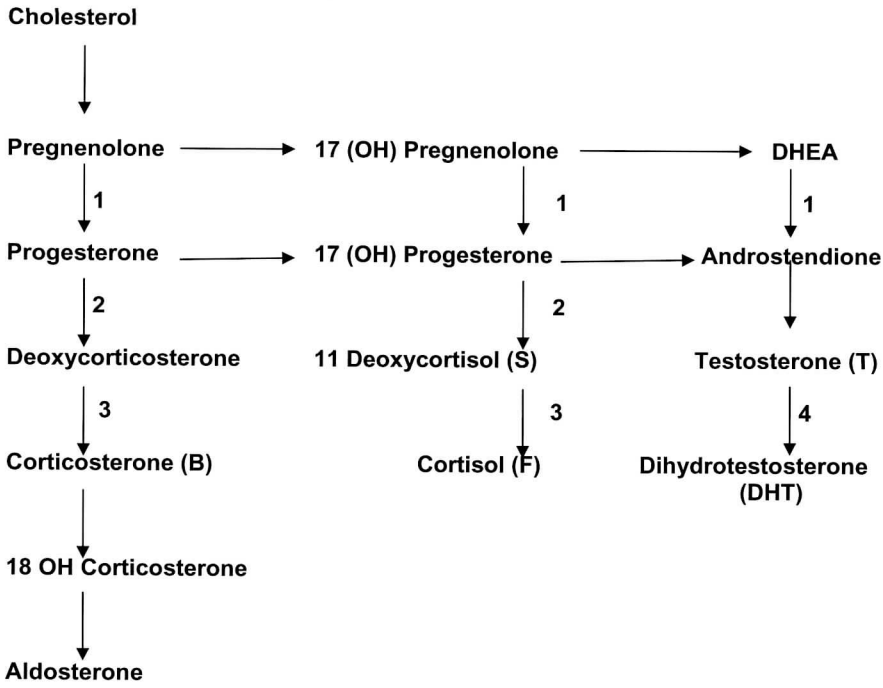


Figure 6: Steroid biosynthesis pathway.



1 : 3 B hydroxyl steroid dehydrogenase
2 : 21 hydroxylase
3 : 11 hydroxylase
4 : 5 a reductase

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NERVOUS SYSTEM & DEVELOPMENT

Diagnosing neurological disorders requires accurate assessment including detailed neurological examination, which is an important step in formulating differential diagnosis and guiding laboratory investigations. Many students, residents, and generalists consider examining the nervous system as one of the most difficult parts of the physical examination. Certain problems frequently face the junior physician including organizing a complete examination in a short time period, and consistently eliciting neurological signs. Certainly, repeated examinations and experience play an important role; however, solid knowledge and use of proper techniques are crucial for eliciting and interpreting neurological signs. In this chapter we present the examination with neuro-anatomical and neuro-physiological outline. Differential diagnosis of some important signs will be presented. Various procedures and techniques of eliciting physical signs and possible pitfalls in the examination will also be discussed.

Organization of the nervous system examination

- Organization is the key to success
- If the examiner has a consistently organized approach to the nervous system examination, he or she will less likely miss important parts of the examination.
- Before proceeding to higher cortical functions, motor, or cranial nerve examination, first assess aspects of the general examination of relevance to the nervous system before conducting the core exam.
- These areas include the following in an ordered approach:

1- Vital signs (including supine and standing blood pressure measurement)
2- Anthropometric measurements (plotted on percentile charts)
2- General examination (skin, skull, spine, dysmorphism, and meningeal signs)
3- Mental status examination (mini-mental status examination)
4- Cranial nerves (I to XII)
5- Motor system (inspection, palpation, and percussion)
6- Cerebellar examination (including gait)
7- Sensory system (peripheral and cortical sensations)

1- VITAL SIGNS

- Temperature:
  High temperature or hypothermia, particularly in infants, may indicate an underlying CNS infection. Temperature instability could be a sign of brain stem dysfunction.
- Blood pressure and pulse:
  Should be measured in the supine and standing positions to assess postural drop as in patients with vaso-vagal syncope. In patients with disturbed level of consciousness or seizures, high blood pressure and bradycardia (Cushing reflex) indicate increased intracranial pressure (e.g. due to hemorrhage or a space occupying lesion).

- Respiration:
  The respiratory pattern may also indicate CNS dysfunction. Cheyne-Stokes breathing (periodic breathing pattern in which phases of hyperpnea regularly alternate with apnea) usually indicates bihemispheric dysfunction. Central hyperventilation (sustained, rapid, and deep hyperpnea) is produced by lesions in the lower midbrain or upper pons. Apneustic breathing (prolonged pause at full inspiration) may occur after damage to the mid or lower pons. Cluster breathing (disordered sequence of breaths with irregular pauses) may result from damage to the lower pons or upper medulla. Finally, ataxic breathing (completely irregular pattern of breathing with random deep and shallow breaths) is usually due to a lesion in the central medulla.

2- ANTHROPOMETRIC MEASUREMENTS

- Weight, height, and head circumference should be measured and plotted on age appropriate percentile charts.
- The distinction between large head (macrocephaly) and large brain (megalencephaly) is important.
- If megalencephaly is suspected the parent's head circumferences should also be measured as benign autosomal dominant megalencephaly is one of the commonest causes.
- Short and tall stature, as well as, under or overweight may be associated with certain disorders and syndromes that may have neurological features or complications as shown in the table.

Abnormality	Syndromes or disease states	CNS features
Short stature	Hypothyroidism Turner's syndrome Cockayne syndrome De Lang syndrome	Mental retardation Hearing loss Peripheral neuropathy Microbrachycephaly
Tall stature	Fragile X syndrome Sotos syndrome Weaver syndrome Marfan syndrome	Mental retardation Macrocephaly Progressive spasticity Risk of embolic stroke
Underweight	Congenital Rubella syndrome Seckel syndrome Rubinstein Taybi syndrome Fetal Hydantoin syndrome	Mental retardation, deafness Microcephaly EEG abnormalities Mental retardation, strabismus
Overweight	Beckwith Weidemann syndrome Bardet Biedl syndrome Prader Willi syndrome	Large fontanels Retinitis pigmentosa Hypotonia

3- GENERAL EXAMINATION

Certain aspects of the general (non-neurological) examination are of relevance to the nervous system including examination of the skin, skull, spine, and examination for dysmorphic features and meningeal irritation signs.

- Skin exam is important as the skin and the nervous system have the same embryologic origin (ectoderm). Therefore, developmental CNS disorders may have associated skin signs (neurocutaneous disorders) as shown in the table.

Neurocutaneous syndrome	Inheritance	Skin manifestations
Neurofibromatosis Type 1	Autosomal dominant	Café-au-lait spots Neurofibromas Axillary or inguinal freckling
Tuberous Sclerosis	Autosomal dominant	Adenoma sebaceum Ash-leaf spots Fibrous plaques Shagreen patches Periungual fibroma
Sturge Weber syndrome	Sporadic	Facial angioma (port-wine stain)
Ataxia Telangiectasia	Autosomal recessive	Telangiectasias (eyes, eyelids, ears, cubital, and popliteal fossas)
Linear Naevus syndrome	Sporadic	Sebaceous naevus Verrucous naevus Acanthosis nigricans

- Examination of the skull for shape, fontanel size and tenseness, sutures for premature fusion or wide separation, and sinus tenderness are important. As well, skull auscultation for bruits may indicate an underlying arteriovenous malformation. For this purpose, the bell of the stethoscope should be placed over the fontanels, eyes, and sides of the head.
- Examination of the spine for deformities (scoliosis, lordosis, gibbus) or midline lesions (defect, hair tuft, or lipoma) may indicate an underlying spinal dysraphism.
- Many syndromes may have associated CNS anomalies or features. It is important therefore to carefully assess the patient for dysmorphic features (face, mouth, palate, hands, and feet).
- Finally examination for meningeal irritation signs is important. Meningeal signs may indicate meningitis, meningoencephalitis, subarachnoid hemorrhage, or cerebellar herniation. Other causes of meningism (symptoms and signs of meningeal irritation with normal CSF findings) include:
    - Local causes: Otitis media, sinusitis, cervical lymphadenitis, apical
    - pneumonia, rheumatoid arthritis
    - Systemic causes: Cerebral malaria, hypernatremic dehydration,
    - bacillary dysentery, typhoid fever
- The following is a description of the signs of meningeal irritation:

    - **Neck Stiffness,** start by active movement. Ask the child to flex his neck as fully as he can and then proceed to passive flexion by gently lifting the head off bed and move it in all directions (flexion, extention, rotation, and lateral direction). Normally the chin could touch the chest without pain). In infants and young children, you should move an attractive object (bright or noisy toy) in different directions to assess the range of neck movements.

- **Kernig's sign,** with the child lying flat on the bed, flex the hip fully and then try to extend the knee slowly. Positive response - if there is resistance or pain to knee straightening in the neck or back indicating meningeal irritation affecting the nerve roots. Pain or spasm in the hamstrings does not represent a positive response. It is used to differentiate between neck stiffness due to meningeal irritation and that caused by local causes such as cervical adenitis.
- **Brudzinski's sign,** flex the neck of the child while observing the hip and knee. It is positive if flexion of the neck results in hip and knee flexion with pain on the back of the neck. All these signs are not reliable in the first 2 years of life because of open fontanels.

4- MENTAL STATUS EXAMINATION

- Detailed assessment may be difficult in young children; however, the mini mental status examination can be done in older children as shown in the table.
- This is a screening test that includes a series of questions and commands to assess various higher cortical functions including orientation, registration, attention, calculation, recall, and language (total score of 30).
- The following is a summary of the mini-mental status examination:

Examination Item	Score
Orientation to time, date, day, month, and year	1 point each
Orientation to place (ward, hospital, district, city, country)	1 point each
Registration (name 3 objects and ask the patient to repeat)	1 point each
Attention and calculation (subtract 7s from 100)	5 points
Recall (repeat the 3 objects named in registration)	1 point each
Language:	
Name 2 objects (e.g. pen and watch)	1 point each
Repeat a sentence	1 point
3 step verbal command	1 point each
1 step written command	1 point
Write a sentence	1 point
Draw intersecting pentagons	1 point

- Cognitive impairment is considered if the total score is less than 23.
- More detailed assessments are needed if it was abnormal.
- May miss subtle or selective cognitive impairment, particularly in executive functions.
- The following are examples on how to inquire about these functions in children:

- **Orientation** (frontal lobe function)
  - What is your name? Do you go to school? Which grade?
  - What is your address? What is your age?
  - Where are you now? What is the time, day, date, month?
  - Ask the younger child to point to his nose, eyes, ear or ask what is this?
- **Attention** (frontal lobe function)

- Young children and infants can be assessed by observing their interest to the surrounding, attention and social contact.
- Forward and backward digits span: Say to the child 5 6 7, then ask him to repeat forward and backward i.e. 7 6 5.
- Note that a 6 years old child can repeat 5 digits forward and 3 digits backward and a 10 years old can repeat 6 digits forward and 4 digits backward.
- Increase the digits up to 7, then recall forward / backward.

- **Thought** (frontal lobe function)
  - Difference between two objects such as chair/ table, shirt/ trousers.
- **Memory** (temporal lobe function)
  - Immediate recall: Give the child name of 3 objects and then ask him to repeat them
  - Short-term memory: Give the child a name of 3 objects (e.g. pen, chair, apple) to remember at the beginning of the interview and then ask him to repeat them at the end.
  - Long-term memory: Ask the child about his address, date of birth and names and ages of his siblings.
- **Calculation** (dominant parietal lobe function)
  - Serial (threes) - subtract 3 out of 10 then 3 out of the remaining.
  - Multiplying (threes) - what is 2 x 3 and what is 2 x the outcome.
- **Spatial** (parietal / posterior temporal / occipital lobe functions)
  - Clock face - draw a clock face and ask the child to fill the hands on a given time
  - Pentagon - draw a five-parted star, diamond or square.
- **Visual & body perception** (parietal / posterior temporal / occipital lobe functions)
  - Agnosia - inability to perceive the sensation despite normal sensory pathways.
  - Ask the child to show you his index finger or thumb (inability to recognize his finger represents finger agnosia)
  - Ask the child to put his right hand on his left ear (inability suggests left or right sided body agnosia)
  - Note that most normal children can identify right and left sidedness in their own body by 6 years and on others body by 9 years.
  - All of these testing rely on intact speech.

5- CRANIAL NERVES

Full cranial nerve examination can be difficult in infants and toddlers. However, simple tests can be applied to evaluate each nerve. At time, observation (inspection) is the only task that can be applied in a young uncooperative child.
Origin of cranial nerves:

Midbrain	III, IV
Pons	V, VI, VII, VIII
Medulla	IX, X, XI, XII

Olfactory nerve (I)

Testing of smell is rarely possible in younger children.

Upper respiratory tract infections are the commonest cause of hyposmia (reduced smell) or anosmia (complete loss of smell).

One should test for smell in older child if any of the following is present:

- Child complains of loss of taste / smell
- Child has evidence of visual field defect
- Frontal lobe trauma (fractures of the cribriform plate, tumor or surgery)
- Hypogonadism / micropenis
- Delayed puberty (Kallman's syndrome)

Instructions**:**

Ask the child to close his eyes and say yes if he smells anything new.

Each nostril should be examined separately (by blocking the other nostril).

Bring the test smell in from the periphery using familiar odors (e.g. mint, vanilla, coffee, chewing gum, orange, toothpaste).

Avoid irritant smell, which will stimulate the 5th cranial nerve (withdrawal).

Optic Nerve (II)

The components to be examined are:

Visual acuity, visual fields, pupillary reaction, fundoscopy, and color vision.

1. Visual acuity:

0 - 5 months:

- Fixate on the mother face (remember human face is the most attractive visual stimulus)
- Fixate and follow a bright object (pen torch or 4 cm red ball). Follow moving object horizontally at distance 100 cm away (90^{o} degree at 6 weeks, 180° degree at 3 months).

6 - 18 months:

- Near vision: Assessed by ability of the child to fix on small objects (size 3-4 mm), e.g. (Smartie or a raisin). Fixation at 6 months suggests adequate vision.
- At 10 months, the child should fix on small objects e.g. decorations as small as 1 mm in size.
- Distant vision: Assessed by the ability of the child to fix on small object at 3 meters. Place a Smartie in front of the child and ask him to get it (if he crawls) or to fix on if he cannot crawl. You can use rolling balls method for assessing near and distant vision

1^{1}/$_{2}$ - 3 years:

- Near vision: Assessed by the ability of the child to fix on a small object.
- Distant vision: Assessed by Sheridan-Gardiner test (letter matching method) at 30 cm. distances, then at 3 meters, or picture book method.

3 - 5 years:

- Near vision/ distant vision: Use Sheridan-Gardiner or picture book for assessing both near and distant vision.

6 years and above:

- Snellen chart should be used. The patient should read the chart from a distance of 20 feet (6 meters), alternately covering one eye, then the other. Pocketsize charts held at 14 inches from the patient's eyes can be used if wall charts are not available. If the patient wears glasses, he should be tested while he is wearing them. The test should be conducted with adequate illumination. If the patient cannot read the largest letter, finger counting and hand motion detection should be performed.

2. Visual field: Again it depends on the age of the child.

- *Old cooperative child:*

Instructions: (**confrontation method**) sit in front of the child so that your head should be in the same level with his head. Instruct him to look at your nose and not to move his head. Now take your hand to the periphery of the visual field with index finger extended and at diagonally opposite position, then tell the child that you are going to flicker your finger and ask him to tell you when he can detect the finger. The moving hand should be at equal distance and should be repeated at different points in the perimeter of the field.

- *Young uncooperative child:*

Move a red ball or torchlight in the visual field from behind to front to see if it attracts his attention.

N.B. Slapping maneuver: wave by your hand as if you are going to slap him is unreliable as the current of air may act as stimulus for blinking.

3. Pupillary reactions:

- Reaction to light: Ask the child to look away from the major source of light in the room and then shine a flash light (pen torch) on the pupil from the side of the eye. Both the pupil on the same side (direct light reflex) as well as the opposite side (consensual light reflex) will constrict (repeat from the other side). Remember that the afferent limb is 2nd and efferent is 3rd cranial nerve.
- Reaction to accommodation reflex: Ask the child to look at a distant object, and then ask him to focus on a finger held close to his nose, the eyes converge and the pupils constrict attempting to look at a close object.
- The interpretation of reflexes:
  1. Loss of direct pupillary response on one side + loss of consensual response on the other side + retention of accommodation reflex is seen in optic nerve lesions. Marcus gun pupil (afferent pupillary defect) results from a lesion in the optic nerve causing slow pupillary dilatation (rather than quick constriction) on the affected side when the consensual reflex is tested.
  2. Loss of both direct and consensual response on the same side + retention of accommodation reflex suggests lesion close to Edinger-Westphal nucleus or ciliary ganglion.

Causes of miosis (small pupil)	Causes of mydriasis (dilated

	pupil)
1. Normal 2. Head injury (pontine hemorrhage) 3. Deep coma 4. Horner's syndrome (sympathetic lesion) 5. Drug toxicity e.g. opium	1. Anxiety & fear 2. Reye's syndrome 3. 3rd cranial nerve lesion 4. Tricyclic antidepressant poisoning 5. Coma due to herniation of brain

4. **Fundoscopy:** To examine the optic disc and retina and should be left to the end
   - Causes of optic atrophy:
     - Birth asphyxia
     - Post papilledema / papillitis
     - Metachromatic leucodystrophy
     - Krabbe leucodystrophy
     - Leigh's encephalopathy
     - Canavan's disease
     - Infantile neuroaxonal dystrophy
   - Causes of optic dysplasia:
     - Idiopathic (isolated)
     - Septooptic dysplasia (isolated or with panhypopituitarism).
5. **Color vision:** Deficiency is commoner in boys (X linked recessive) The test can be done at 8 years of age by the ophthalmologist.

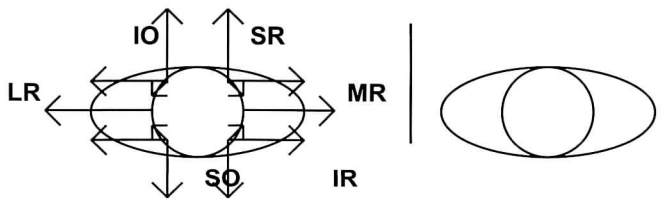
Occulomotor, Trochlear, and Abducent nerves (III, IV, VI)

Items to evaluate:
- Position of the eyes at rest (squint)
- Conjugate eye movements
- Eye movements in relation to head and body movements.

•Instructions:
- *Older child*:

Ask the child to follow an object (e.g. your finger) in different directions and then ask him how many fingers or objects he has seen (diplopia).
- *Infant / young child:*

Attract the attention of the child by a small toy and move it in vertical, horizontal and oblique directions.



IO = Inferior oblique muscle
SR = Superior rectus muscle
SO =Superior oblique muscle
LR = Lateral rectus muscle
MR = Medial rectus muscle
IR = Inferior rectus muscle

NB. Remember that the rectus muscles move the eyes in the direction of their names (e.g. the medial rectus moves the eye medially). The oblique

muscles move the eyes in the opposite direction (e.g. the superior oblique moves the eye inferiorly). As well, the superior oblique muscle moves the eye inward and the inferior oblique muscle moves the eye outward in a rotatory movement.

Causes of ocular nerve palsy:

- Cerebral palsy
- Post meningitis
- Guillain-Barré syndrome
- Brain stem tumor
- Raised ICP (false localizing sign)

Oculomotor nerve (III)

All extra ocular muscles are supplied by oculomotor nerve except superior oblique (4th nerve) & lateral rectus (6th nerve).

Palsy results in the following features:

- A paralytic divergent squint
- The affected eye is deviated downward and outward with impairment of adduction and elevation
- Dilatation of the pupil (normal in nuclear lesion)
- Ipsilateral ptosis
- Diplopia in all directions except lateral gaze to the side of the lesion

Trochlear nerve (IV)

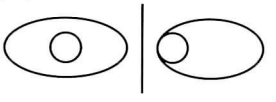
Supplies the superior oblique muscle. Palsy results in:

- Compensatory torticollis (head tilt)
- No obvious squint, it is only manifested on looking downward and inward.
- Diplopia occurs on attempted down gaze.

Abducent nerve (VI)

Supplies the lateral rectus muscle. Palsy results in:

- Convergent squint (medial deviation of the eye)
- Inability to abduct the eye beyond midline
- Compensatory torticollis
- Failure of lateral gaze on the affected side.



Trigeminal nerve (V)

Motor component:

Motor function may be tested by examination of masseters, pterygoids and temporalis muscles during mastication as well as by evaluation of the jaw jerk.

- **Instructions:**

- *Older child*: Ask the child to open and close his mouth, if one side is paralyzed the jaw will be deviated to the paralyzed side when the mouth is opened (pterygoids). Ask the child to clench his teeth, then palpate for masseter and temporalis muscles.
- *Infant and young child*: Put a wooden spatula in the child's mouth and let him bite it. With normal pterygoids he will resists its removal. Or

offer a nipple or bottle and observe for sucking (strong sucking suggests normal masseters).

Jaw reflex (5^{th} nerve constitutes both afferent and efferent limbs)

- Ask the child to open his mouth a little, place your forefinger over the chin and then tap your forefinger in a downward direction with a tendon hammer. Normally there is a weak or absent jerks of the jaw. Exaggerated jerk (brisk jaw closure) is seen in patients with frontal lobe dysfunction (release reflex), or in extrapyramidal disorders (e.g. Parkinson's disease).

Sensory component:

Test touch and pain sensations over the 3 sensory divisions: ophthalmic, maxillary, mandibular.

• Instructions:

- *Older child*: Touch the face in three different regions with a wisp of cotton (not rubbing or dragging) over the forehead (ophthalmic), cheeks (maxillary), and chin (mandibular). Ask the child to close his eyes and say yes if he feels the touch.
- *Neonate and infant*: Elicit rooting reflex, glabellar reflex

Corneal reflex (afferent V1 and efferent VII)

• Particularly in patients with disturbed level of consciousness

- *Older child*: Ask the child to look in one direction and approach the cornea from the opposite side with a wisp of cotton, observe for the blink.
- *Infant and young child*: Blow air gently and observe for eye blink.

Facial nerve (VII)

- Observation:
- Facial asymmetry (absence of frontal wrinkles)
- Flattening of nasolabial folds.
- Open eye (widened palpebral fissure)
- Deviation of the angle of the mouth (drooping of the corner of the mouth).
- Instructions:
- *Older child:* Ask the child to raise his eyebrows, to frown, or to look at your finger held above the head (occipito-frontalis muscle).
  - Ask the child to close his eyes tightly while you are trying to open them (orbicularis oculi).
  - Ask the child to smile or to show his teeth (levator labii muscle).
  - Ask the child to whistle (orbicularis oris).
  - Ask the child to puff out his cheeks and to keep them out while you are tapping with your finger over both cheeks to detect ease of air expulsion on the affected side (buccinator).
- *Infants and young child*: No particular maneuver is used just observe the child's face while he is crying or laughing. Lower motor neuron lesion tends to equally involve upper and lower facial muscles while upper motor neuron lesion affects the movements of the lower part of the face.

Sensory: Chorda tympani supplies taste to the anterior two-thirds of the tongue.

Examination of taste is hard to perform and not usually recommended.
* *Older cooperative child:* Can be tested by placing solution of saline or glucose on one side of the protruded tongue (normal child can identify the substance with little difficulty).

Vestibulocochlear nerve (VIII)

A) Cochlear division:

Test for hearing according to the age. Accurate assessment can be difficult, as it requires time, expertise and quite environment.

- *Neonate:* Startle or quieted to a loud stimulus (a clap)
- *6 - 8 months:* Distraction test
- *Older child:* Audiometry, Rinne's test, Weber's test

One should test for hearing if any of the following is present:

- Family history of deafness
- Parents are concerned that the child is deaf
- Post meningitis
- Cleft palate
- Cerebral palsy
- Low birth weight
- Delayed speech

B) Vestibular division:

This cannot be effectively tested at bedside. It is usually tested along with cerebellar function in order to assess balance and gait.

Clue to vestibular dysfunction:

- Jerky nystagmus
- Disturbed balance and posture
- Nausea and vertigo with change of position
- Caloric stimulation test.

Glossopharyngeal and Vagus nerves (IX, X)

The IX and X nerves are usually considered together as they have the same exit from the skull and run a similar course together. The glossopharyngeal carries common sensation from the pharynx, tonsil, soft palate, posterior $^1/_3$ of tongue and taste sensation from posterior $^1/_3$ of tongue. While vagus nerve gives motor fibers to pharyngeal, palatal muscles and vocal cord.

Clue to dysfunction:

- Dysphagia with drooling and choking
- Nasal regurgitation during swallowing of liquid due to palatal weakness
- Nasal speech, hoarseness of voice, bovine cough
- N.B. Unilateral IX - X nerve palsy is asymptomatic except for hoarseness
- of voice while bilateral palsy results in the above symptoms

- Instructions:
  - *Older child*: Talk to the child to comment on his voice (hoarseness of voice) then ask him to cough (bovine cough). Ask the child to open his mouth and observe the position of the uvula at rest, then ask him to say " Aah " and observe the movement of uvula during phonation. Unilateral nerve palsy causes deviation of uvula to the normal side.

Palatal reflex: Touch the child's soft palate with tongue depressor; it will lead to elevation of soft palate & retraction of uvula.
Gag reflex: Touch the child pharyngeal wall with spatula; it will stimulate gagging (elevation of the pharynx & tongue retraction).
N.B. This reflex should not be elicited in a conscious child. In both reflexes the afferent is IX nerve and the efferent is X.
* *Infant:* Observe the movement of the palate, observe the act of sucking (V, VII, IX, XII), observe the act of swallowing (IX, X, XI), observe for regurgitation, choking during sucking and swallowing and observe the cry.

Spinal accessory nerve (XI)

Muscles supplied by the accessory nerve are trapezius & sternocleidomastoid.

- Instructions:
- *Older child:* Ask the child to shrug his shoulder (this tests the upper part of trapezius) weakness of trapezius results in ipsilateral dropping of the shoulder. Put your hand on the medial side of the child jaw and ask him to push against your hand while you are palpating the opposite sternocleidomastoid muscles. Put your hand on the forehead of the child and ask him to flex his head forward against the resistance of your hand, compare the 2 sternocleidomastoid muscles.
- *Infant:* Let the mother throw a toy beside him while he is looking at it feel the sternocleidomastoid muscle of opposite side or let his mother call his name and when he turns his head to her feel the sternomastoid muscle of opposite side.

Hypoglossal nerve (XII)

- Look at the child's tongue, while inside the mouth observe for size, position wasting, fasciculation (sign of lower motor neuron lesion).
- This is true because of the absence of subcutaneous fat.
- Instructions:
- Ask the child to stick his tongue out, observe for any deviation to either side.
- Unilateral paralysis causes the tongue to deviation towards the weak side
- Ask the child to push by his tongue against tongue blade (on each side).

6- MOTOR SYSTEM

The motor system includes the assessment of the following:

- **Inspection** (posture, gait, muscle bulk, involuntary movements)
- **Palpation** (tone, power)
- **Percussion** (reflexes)

Muscle bulk:

Notice the presence of atrophy or hypertrophy and whether it is generalized / localized, proximal / distal and symmetrical / asymmetrical.
Atrophy (wasting) of muscles:
Muscle atrophy implies lower motor neuron lesions, muscle disease or prolonged immobilization.
N.B. Proximal wasting and weakness indicates myopathy (except myotonic dystrophy which is distal), while distalweakness indicates

peripheral neuropathy (except spinal muscular atrophy which is proximal).

Hypertrophy & pseudohypertrophy:

Muscle hypertrophy can be present in a muscular dystrophy, e.g. Duchenne muscular dystrophy, autosomal recessive muscular dystrophy, myotonia congenita and congenital hemihypertrophy

Involuntary movements:

There are many types of involuntary movements. Good description of them is more important than the exact diagnosis.

Chorea: Rapid, jerky, intermittent, random, non-stereotypic, dancing movements that usually affects the proximal extremities and face, but also can involve the distal limbs. They are aggravated by activity and emotional stress, and disappear during sleep.

Athetosis: Slow, continuous, writhing movements of the extremities and face, particularly the proximal parts, that is usually combined with chorea (choreoathetosis). They are aggravated by voluntary activity and emotional stimuli.

Tic: Complex repetitive movements. They get worse when the others are watching, and usually affect eye muscles, facial muscles, upper limbs. They don't interfere with the child's normal activity and disappear during sleep.

Tremor: Rhythmical alternating movements, they may occur at rest or action. It can affect the hands or neck and head (titubation)

Myoclonus: Sudden, quick, jerky (shock-like) movement of a limb due to contraction of a group of muscles.

Dystonia: Slow, twisting, continuous contraction of both agonist and antagonist muscles resulting in a sustained limb posture (e.g. writer's cramp).

Fasciculation: Rapid involuntary contraction of one or more muscle fiber, which may leaves furrows on the skin overlying the muscle. It occurs in lower motor neuron lesions (e.g. Werdnig-Hoffman's disease, poliomyelitis, neuropathy). Best seen in the tongue and thenar eminence where there is minimal subcutaneous fat.

Muscle tone:

Defined as the resistance (tension) in the muscle to passive movement. Testing the tone: Start by observing the position of the limbs. Frog like position (hypotonia), scissoring (spasticity), or dystonic posture.

Upper limbs:

- Shake test: (Test for range of movements around the joint) Hold both wrists of the child in your hand and quickly shake his hands to and fro around wrist joints, watch how freely the hands waggle.
- Resistance to passive stretch around the joints:
    - Wrist - Flexion / extension
    - Elbow - Flexion / extension, supination / pronation.
    - Shoulder - Flexion / extension, abduction / adduction, internal rotation /external rotation
- N.B. In normal term infant if the upper limb is pulled gently across the chest, the elbow normally does not extend beyond the midline. The elbow of hypotonic child extends beyond the midline with ease (*Scarf* sign).

- Palpation of the muscle for floppiness and consistency.

Lower Limbs:

- Shake test: Hold both legs of the child and shake both feet to and fro around the ankle joints, watch how freely the feet waggle.
- Resistance to passive stretch around the joints:
    - Ankle (Dorsiflexion / plantar flexion, inversion / eversion)
    - Knee (Flexion/ extension, popliteal angle)
    - Hip (Abduction / adduction, flexion / extension, internal rotation / external rotation)
- Comment on the tone; whether it is normal or decreased (hypotonia) or increased (hypertonia) which is either spasticity or rigidity:

Spasticity: Which means increase of resistance of the muscle to passive movements with maximum resistance at the beginning of movement.

- It is due to pyramidal tract lesion.
- It is usually associated with increased reflexes.
- It does not change with position or emotional status.

Rigidity: Which means increase of resistance of the muscle to passive movements which is uniform throughout the range of movement which either smooth (lead pipe) or intermittent (cog wheel)

- It is due to extra-pyramidal lesion.
- It is usually associated with normal reflexes
- It changes with the position or emotional status

N.B. The resistance to passive movements is affected by muscle status and bony deformity.

Muscle power:

Power is graded according to MRC grading scale into:

0 No contraction
1 Flicker of contraction
2 Active movement with gravity eliminated
3 Active movement against gravity
4 Movement against moderate resistance
5 Normal power

Golden rules in assessing the muscle power in children:

- Have a simple system rather than an exhausting one.
- First demonstrate to the child the action you want him to do.

Upper limbs:

- *Older children*:
    - Ask the child to abduct his arms so that they are about 45° away from his body then ask him to push his elbow away from his body while you are pushing against it. (test for shoulder abduction C5), then ask him to push his elbow into his body while you are pushing against it (test for shoulder adduction C7).
    - Ask the child to bend his arm so that the elbow is at right angle then ask the child to flex his forearm while you are pushing against it (test for elbow flexion C5, 6), then ask the child to extend his forearm while you are pushing against it (test for elbow extension C7, 8).
    - Ask the child to make a fist (C8).
    - Ask the child to extend & flex his wrist while you are pushing against it (test for wrist flexors and extensor C6, 7).

- Ask the child to spread out his fingers (interossei muscle T1), then to bring his fingers together (lumbricals T1).

- *Infants and younger children*:
  - It is difficult to perform formal test for muscle power at younger children and infants. However some maneuvers can be used.
  - Shoulder girdle muscles: During vertical suspension of the child observe for slipping through the hands. If no slipping suggests strong shoulder flexors and adductors, offer object (toy) above the level of the child's head and let him to pick it or let him to throw a ball or comb his hair.
  - Elbow flexors group: While pulling the infant up from supine to sitting position, observe for flexion at the elbow with resistance to pull anticipation reflex). Flexion of elbow with resistance to pull suggests strong biceps muscle.
  - Hand grip muscles: Offer an object or put it in the child hand and observe how the child grips it.

Lower Limbs:

- *For older children:*
  - Ask the child to lift one leg straight off the bed, to keep it up while you are pushing down against him (test for hip flexion L1, 2).
  - Ask the child to press the leg straight down into the bed while you are pulling up against him (test for hip extension L5, S1)
  - Ask the child to bend his knee to a right angle, then ask him to straighten his knee while you are pushing against it (test for knee extension L3, 4).
  - Ask the child to keep his knee at right angle and to flex his knee fully while you are pulling against him (test for knee flexion S1).
  - Place your hand on the dorsum of the child foot and ask him to pull his foot toward him while you are pushing against him (ankle dorsiflexion S1, S2).
  - Place your hand on the sole of the child foot, ask him to push down while you are pulling against it (Ankle planter flexion L4).
  - Ask the child to bend his big toe up and down against resistance (L5).
- *Infant and younger children*:
  - If the child is moving his limb spontaneously, it suggests that the muscle power of the limb is grade 3 or more.
  - Put the lower limb of the child in a position of flexion and adduction at hip joint with flexion at the knee joint and observe if the child can hold his leg in this position or not. Holding in this position suggests strong hip flexors and adductors
  - Hold the child's thigh and tickle the child at the sternum or axilla to see if he can flex or extend his knee (suggests normal knee flexors / extensors).
  - Hold the child's leg and tickle him, to assess the movement around the ankle.
  - N.B. You can apply resistance to the limb while it is moving to assess power.

Shoulder girdle: Ask the child to do the following:

- To comb his hair or to throw a ball or to lift his hands up.

Pelvic girdle: Ask the child to do the following:

- To walk, to run, to hop, to jump, to get up from sitting or squatting position, to climb stairs, demonstrate the Gower's sign, Trendelenburg's sign.

Trunk muscles: Observe the following:

- *Older child*:
  - Ask him to sit up from supine position with hands folded across the chest, ability to sit without aid suggests strong trunk muscles. (Inability to do so means weakness of T1-12).
- *Infants:*
  - Observe the child while he is sitting from supine position, then observe him pivoting.
  - While the child is sitting with his head central give sideways push and watch if he falls sideways or depend on his hands for lateral support

N.B. *Examination of the back is important and essential in assessing the lower limb. Start the examination of the lower limbs by gait assessment if the child's condition permits.*

Reflexes:

GOLDEN RULES:

- **The reflexes should be assessed at the end of the neurological examination as striking the child with a tendon hammer is a frightening thing.**
- **Don't try to elicit a reflex while the child is moving that limb.**
- **Try to distract the child's attention by talking to him while eliciting the reflex.**
- **Hold the hammer by the end of the handle so be a swinger not a stubber when striking a tendon.**
- **Expose the muscle suppose to contract by the elicited reflex**
- **An absent reflex is not absent, but simply not elicited; proceed to reinforcement.**
- **Reflexes are rarely absent or pathologically brisk in the absence of other neurological abnormalities.**
- **The head should be central when testing reflexes as the briskness of reflexes may be increased on the side to which the face is turned. (asymmetric tonic neck reflex).**
- **Children tend to have brisker reflexes in their legs than their arms.**
- **Absence of ankle reflex alone may be the first evidence of a peripheral neuropathy, while absence of all reflexes except the ankle reflex is more common in myopathy.**

Summary of the deep tendon reflexes:

Deep tendon reflex	Muscle involved	Nerve supply	Root supply
Biceps	Biceps	Musculocutaneous	C5, C6
Triceps	Triceps	Radial	C6, C7, C8
Pectoralis	Pectoralis Major	Pectoral	C6, C7, C8
Brachioradialis	Brachioradialis	Radial	C5, C6
Finger flexors	Flexor Digitorum	Median and Ulnar	C7, C8, T1
Knee	Quadriceps Femoris	Femoral	L2, L3, L4
Adductor	Adductors	Obturator	L2, L3, L4
Ankle	Soleus / Gastrocnemius	Sciatic / Tibial	S1, S2
Planter	Small foot muscles	Plantar	L5, S1, S2

Reflexes of the upper limbs:

Biceps reflex: (Musculocutaneous nerve, C5,6)
Semiflex the child's arm at the elbow, put your thumb over the radial insertion of the biceps, then strikes the tendon with a hammer.
Response: flexion of elbow with contraction of the biceps muscle

Triceps reflex: (Radial nerve, C6, 7, 8)
Flex the child's arm at elbow, and then tap on the triceps tendon above the elbow.
Response: extension of elbow with contraction of the triceps muscle.

Brachioradialis (Supinator) reflex: (Radial nerve, C5, 6)
With child's arm half flexed - half pronated, tap over the lower end of radius.
Response: flexion of elbow with contraction of Brachioradialis muscle

N.B. If you could not elicit upper limb reflexes do **reinforcement procedure** by asking the child to clench his teeth while striking the tendons.

Reflexes of the lower limbs:

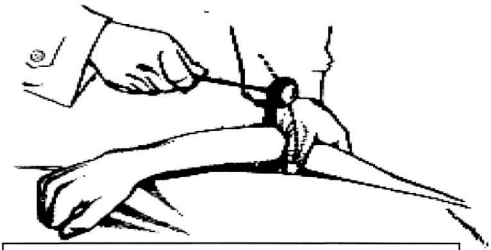
Knee jerk: (Femoral nerve, L2,3,4). Flex both the knees together at right angle by placing your left arm under them or let the child be seated on the edge of the bed with his legs hanging down, then strike the patellar tendon with the hammer.
Response: extension of the knee with contraction of the quadriceps.
Note that preterm infants, particularly those less than 33 weeks of gestation, have decreased elicitation rates for patellar and biceps reflexes and have decreased overall reflex intensity when compared with their older counter parts.

Ankle jerk: (Sciatic / Tibial nerve, S1,2). Rotate the patient's leg externally and flex the knee at right angle and with gentle traction dorsiflex the foot, then strike the tendoachillis.
Response: planter dorsiflexion of the foot with contraction of the gastrocnemius muscle.

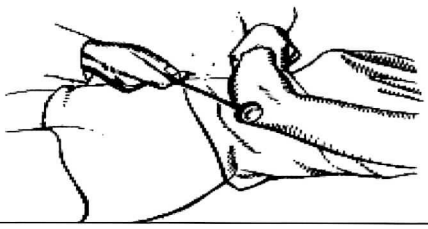
If the reflexes of lower limb are not elicited do **reinforcement procedure** by asking the child to interlock his fingers in a "monkey grip" and pulling it hard just before the hammer strikes the tendon.

Deep tendon reflexes are graded as follows:

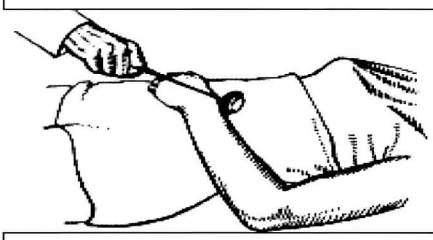
0	=	Absent
1 +	=	Weak response (present)
2 +	=	Normal response
3 +	=	Brisk response
4 +	=	Pathological hyperreflexia



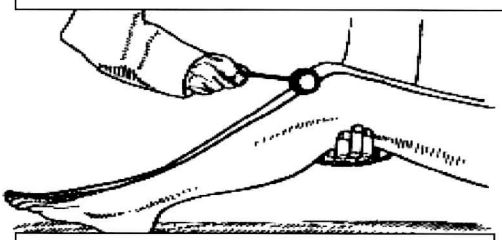
Eliciting the biceps Jerk, C.5



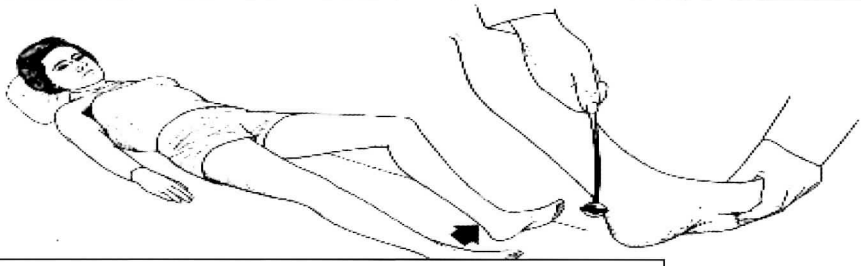
Eliciting the triceps Jerk, C.6, C7



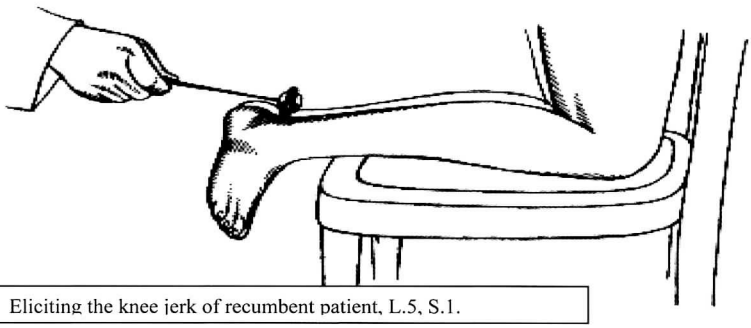
Eliciting the Supinator jerk, (C.5), C.6.



Eliciting the knee jerk, L.3, L.4.



Eliciting the knee jerk of recumbent patient, L.5, S.1.



Eliciting the knee jerk of recumbent patient, L.5, S.1.

* From Macleod, Monro, Clinical examination, 7th edition

Characteristics of pathological hyperreflexia (+4):

Exaggerated reflexes are considered pathological (+4) if associated with spasticity, clonus, or reflex spread (e.g. hip adduction on tapping the knee jerk or finger flexion on tapping the biceps).

Clonus:

Clonus is a rhythmic series of involuntary muscular contractions evoked by a sudden sustained stretching of tendon. Presence of clonus indicates pyramidal tract lesion.

Ankle clonus: To elicit it, keep the child in supine position with his knee flexed by one of your hands while the other hand holds the forefoot then dorsiflex the forefoot with a quick and sustained push. Positive response: the foot goes through plantar flexion and dorsiflexion movements repeatedly > 3 beats.

N.B. Up to 10 beats of ankle clonus may be normal in the first 2 months of life, after this age ≥ 4 beats is pathological.

Patellar clonus: To elicit it, hold the superior border of the patella give a quick and sustained push towards the foot while the child is lying supine in bed with his knee extended. Positive response: patella moves up and down repeatedly.

Superficial reflexes:

Plantar reflex (L4, S1)

- Stimulate the sole of the foot along the outer border of the foot with a blunt instrument e.g. handle of hammer, key, thumb in neonates and infants. Start near the heel and carry the stimulus over the base of metatarsals ending near the ball of the great toe.
  - Normal response: Plantar flexion of the big toe with the flexion to other toes or withdrawal of the entire leg in the child.
  - Abnormal response: Dorsiflexion of the big toe at metatarso-phalangeal joint with sluggish spreading or fanning of the other four toes. (Positive Babinski sign)
- N.B. The plantar response is normally extensor in all infants in the first 6 months of life and in 75% of infants up to the age of 1 year.

Abdominal reflexes (T8-12)

- Strike the four quadrants of the abdominal wall with a blunt instrument from outwards to inwards while the child is lying flat and relaxed.
  - Normal response: Contraction of the abdominal wall at the stimulated quadrant and deviation of the umbilicus to the same side.
  - Abnormal response: No contraction with exaggerated deep tendon reflexes suggesting pyramidal tract lesion.

Ano-cutaneous reflex (Anal wink S2, 3, 4)

- It is elicited by stimulation of the perianal skin by pinprick.
  - Normal response: Contraction of external anal sphincter with indrowing of the anal opening.
  - Abnormal response: Loss of the reflex means lesion of S2, 3, 4.

Cremasteric reflex (L1, 2)

- It is elicited after 10 days of life in male child. It is tested by striking the upper medial aspect of the thigh.
    - Normal response: Elevation of the scrotum of the same side of stimulation due to contraction of the cremasteric muscle.
    - Abnormal response: No elevation of the scrotum means a lesion above L2

7- CEREBELLUM & COORDINATION:

It is affected by poor visual acuity, cerebellar disorder and sensory loss. The maneuver used to assess the coordination depends on the age of the child and presence or absence of muscle weakness.

Upper Limbs:

- *Older child:*

Finger-nose test:

- Ask the child to place the tip of the index on his own nose starting from full abduction of the arm with eyes open, then with eyes closed. Observe for intention tremor and dysmetria (overshooting).

Finger to finger nose test:

- Hold your finger at arm's length in front of the child, ask him to touch your finger with his index finger then touch his nose, move the target finger, (your finger) in different directions. Ask him to repeat faster if he did it correctly.
- Both intention tremor and overshooting indicates cerebellar lesion.

Dysdiadochokinesia:

- Ask the child to put one hand on the back of the other quickly and regularly (always demonstrate for the child)
- Ask the patient to tap the back of his right hand alternately with the palm of his left hand and vice versa.
- Ask the child to rapidly supinate and pronate his hands together; observe for symmetry and regularity.
- Ask the child to do the following:
    - To mimic playing piano
    - To touch the finger of the hand alternatively with the thumb of the same hand (finger opposition)
    - To button a shirt
    - To write his name
- N.B. Children above 5 years should do these tests easily.

- *Infant and younger child*: Cannot cooperate to perform formal testing. Offer a toy and observe the child's approach looking for dysmetria or intention tremor. Offer cubes and observe him while building them. Ask the child to put a pen into a pen-top held by you.

Lower limbs:

Heel-shin test:

- While the is child is lying down ask the child to place his heel on the opposite knee and then run it downwards over the shin of the tibia to the foot, observe if he can follow the line of the tibia smoothly and accurately.
- This maneuver should be done with eyes opened and repeated with eyes closed to differentiate between sensory and cerebellar ataxia.

Toe finger test:
- Ask the child to lift his big toe and touch your finger with it, observe for dysmetria and intention tremor.

Tap test:
- Ask the child to tap his foot against your hand, observe for dysmetria and intention tremor

Gait:
- Ask the child to walk (unsteady gait)
- Ask him to hop (if he is older than 4 years)

Tandom - walk test:
- Ask the child to walk on straight line with heel to forefoot for a distance more than 8 steps.

Trunk:
- *Older child*:
  - Ask the child to sit up with hands folded across his chest, when he sits up give sideways push to see if he falls sideways or not.
- *Younger child:*
  - While the child is sitting observe for swaying movement of trunk, nodding and bobbing of head, then move an interesting toy around 180° and see if the child is unable to turn to the toy except by supporting himself on his hands or by altering his sitting position.

8- SENSORY SYSTEM
- The sensory system is very difficult to examine in children who may not cooperate. Young children are easily frightened, distracted and fatigued. Therefore patience is needed.

Golden Rules:

- **Test each dermatome separately**
- **Start by deep sensation e.g. joint position as they are quick and require little concentration, then touch, followed by pain using a blunt pin to avoid frightening the child.**
- **Start by area you expect to be affected, then work towards area of normal sensation. In all parts of sensory testing it is essential first to teach the child about the test.**
- **Check that the child has understood and carried out the test appropriately.**
- **Some tests are time consuming and you may need to repeat them in another sitting.**
- **The items to be tested are: Superficial sensation (touch, pain, temperature; Deep sensation (Joint position, vibration, pressure); Cortical sensation (steriognosis, two-ponit discrimination, extinction).**

A. Deep sensation:

Sense of movement (joint position):
- Upper limbs: Hold the middle phalanx of the child's index finger by side (not the pulp), then flex and extend the distal phalanx telling the child which direction you mean by up and down, then repeat it with his eyes closed.

- Lower Limbs: Use the same idea with the big toe holding terminal phalanx by the sides.

Vibration Sense:

- It is difficult and unlikely to give more information than the joint position sense.

B. Superficial Sensation:

Equipment: Cotton ball, safety pin, tube with warm & cold water (ice cubes)

Light touch:

- Use a wisp of cotton and touch the skin lightly (don't rub), then ask the child to say "yes" if he can feel it with his eyes closed. Examine all dermatomes (Fig. 2)

Pain sensation:

- To test for pain, a similar approach is used with stimulus being the tip of a pin.
- The child should be instructed to respond by saying "sharp" or "dull" and should not just say "yes" every time he feels. This is to avoid the child's response to the pressure aspect of the pinprick. Younger child or infant responds by facial expression and withdrawal of limb.

Temperature sensation:

- Use two tubes of cold and warm water.
- Demonstrate to the child that you are going to touch him with either warm or cold tube, and then he should tell it if it is "cold" or "warm".

C. Cortical sensation:

- Result from an upper motor neuron lesion usually involving the right parietal lobe.

Astereognosis:

Failure to recognize familiar objects when put on the hand with closed eyes (e.g. a coin or key in the hand).

*A*graphesthesia*:*

Failure to recognize numbers & letters when written in the palm while the eyes are closed.

2 point discrimination:

Ability to identify 2 points applied simultaneously to the dorsum of the foot or pulp of the finger. Approximate the two points together until he perceives them as one point

Tactile localization:

Inability to localize the point of touch with closed eyes.

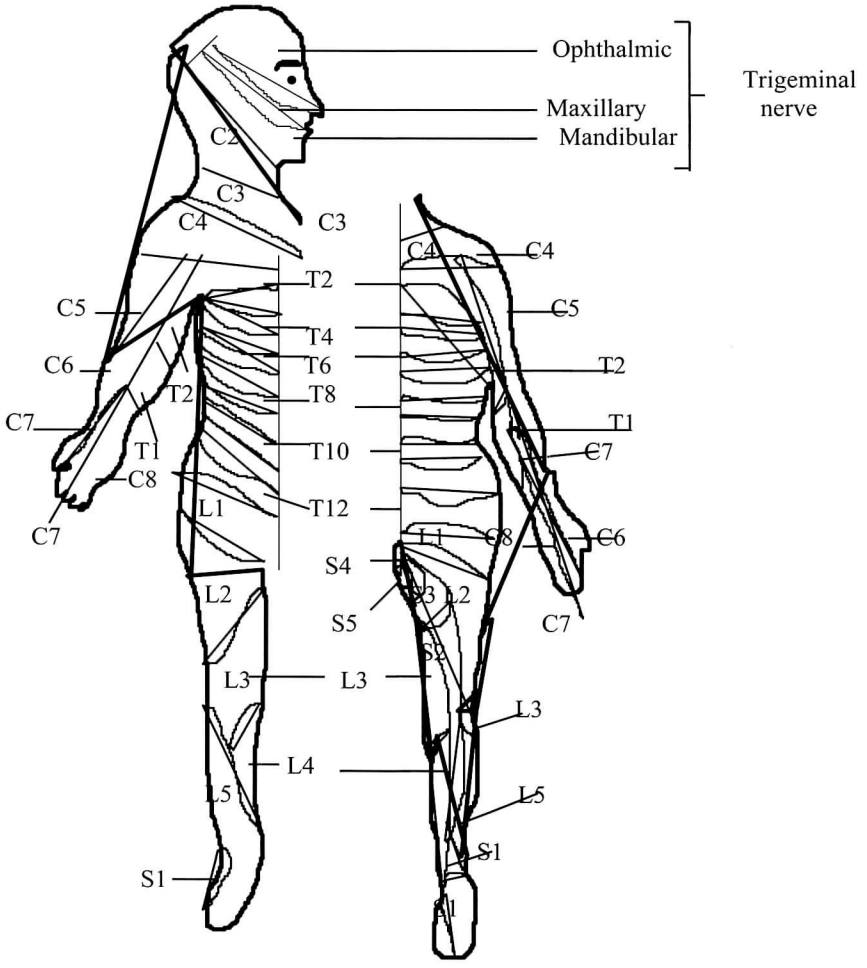


Fig. 2 Distribution of dermatomes

COMMON PAEDIATRIC NEUROLOGICAL DISORDERS

ABNORMAL HEAD SHAPE OR SIZE

Abnormal head shape:

Inspection of the shape of the head is best done when the child is sitting up on the mother's lap and look from all angles.

Abnormal shapes of the head:

1. Scaphocephaly (dolechocephaly) - Increased anterioposterior diameter due to premature closure of sagittal suture.
2. Brachycephaly - Increased transverse diameter due to premature closure of both coronal sutures.
3. Plagiocephaly - Asymmetrical head due to premature unilateral coronal or lambdoid suture closure.
4. Trigonocephaly - Narrow pointed forehead with hypotelorism due to premature closure of metopic suture.
5. Oxycephaly (acrocephaly, turricephaly) - High narrow tower shaped head due to premature closure of all sutures.

Abnormal head size:

1. Measure the head circumference (3 times) around the most prominent part of the head to get the maximum head circumference.
2. Measure the head circumference of the parents.
3. Plot on percentile chart (correct for prematurity in the first 2 years of life).
4. Always compare with previous head circumference and percentile charts.

Average normal head circumferences:

35 cm at birth
47 cm at 12 months (i.e. 12 cm in 12 months)
49 cm at 2 years (i.e. 2 cm in 2nd year)
50 cm at 3 years
52 cm at 6 years
53 cm at 10 years
56 cm at adult

- Disorders of cranial size are either small head (microcephaly) or large head (macrocephaly).
- Large head is defined as head size above two standard deviation of the normal it could be due to big brain (megalencephaly) or bone disorder.
- Small head is defined as head size below two standard deviation of the normal. It could be due to small brain (micrenencephaly) or craniosynostosis.

Clues to craniosynostosis:

- Presence of ridges along the suture lines.
- Abnormal shape of the head depending on the involved sutures.
- Abnormal eye finding (proptosis).

Clinical approach to abnormal head size:

History:

When did the parents observe abnormality of head size? Ask about the gestational age, history of trauma, symptoms suggestive of raised ICP, history suggestive of congenital infections, birth asphyxia, drug intake or X-ray exposure during pregnancy, history of encephalitis or meningitis, and family history of small or large head.

Examination:

A*) Inspection*:

- Local, comment on the shape and head movements. Look for signs of hydrocephalus (large fontanels, presence of prominent scalp veins, sunset sign, shiny skin of the head)
- General, growth parameters (short in achondroplasia, tall in Sotos syndrome). Examine the child's alertness and look for neurocutaneus stigmata and dysmorphic features suggesting a particular syndrome.

B) *Palpation*:

- Palpate the head for sutures, ridges, fontanels, and shunts.

C) *Auscultation*:

- Listen for bruit on temporal region, over the eye balls, and retroauricular region. Then proceed to:
  - Eye examination: visual acuity, visual field, ocular movements, pupil, fundus, sunset sign, squint nystagmus.
  - Back examination: spina bifida, scoliosis
  - Lower limbs examination: signs of UMNL e.g. hydrocephalus, LMNL e.g. spina bifida, leucodystrophy, cerebellar signs e.g. Dandy Walker cyst
  - Abdominal examination: For hepatosplenomegaly (storage disease, mucopolysaccharidosis, congenital infections)
- N.B. In neurological evaluation of a child with large head start by lower limbs as their tracts are closer to the ventricles.

Causes of microcephaly:

A) *Primary* (congenital)

- Chromosomal (trisomy 18), syndrome (Aicardi syndrome, Rubinstein syndrome, Angleman syndrome), inherited (autosomal recessive, autosomal dominant, X-linked recessive).

B) *Secondary* (acquired)

- Infections (e.g. TORCH), fetal alcohol syndrome, hypoxic ischaemic encephalopathy, encephalitis, meningitis, head injury, metabolic (e.g. phenylketonuria), endocrine, (e.g. hypothyroidism).

Causes of macrocephaly: (remember by 5L)

1. Large skull
   - Achondroplasia
   - Rickets
   - Osteogenesis imperfecta
   - Chronic haemolytic anemia

- Cleidocranial dysostosis

2. Large brain
   a) Generalized
      - Familial (Autosomal dominant)
      - Sotos syndrome
      - Neurocutaneous syndromes
      - Storage diseases:
        - Lipidosis (Tay-Sachs)
        - Mucopolysaccharidosis
        - Leucodystrophy (Alexander, Canavan)
   b) Localized
      - Cerebral tumor
      - Arachnoid cyst
3. Large ventricles / subarachnoid spaces
      - Hydrocephalus
5. Large bleed - chronic subdural haematoma
      - Traumatic
      - Post meningitis

SUDDEN ONSET OF UNSTEADY GAIT (ATAXIA)

Clinical approach:

History:

Trauma, contact with or previous history of infectious diseases (e.g. chicken pox or mumps), symptoms of raised ICP, drug ingestion (antihistamines, anticonvulsants, antidepressants), bleeding disorders, recurrent infections, and family history of migraine or neurodegenerative disease.

Examination:

- Examine for *cerebellar signs.*
  - Speech, nystagmus, intention tremor, dysmetria, dysdiadochokinesia
  - Then ask the child to sit, walk, run to demonstrate ataxic gait.
- Examine for the *cranial nerves* involvement indicating brain stem pathology
- Examine the *deep tendon reflexes.*
  - Pendular reflex suggests cerebellar pathology.
  - Exaggerated reflexes in four limbs suggest brain stem pathology, while exaggerated reflexes in lower limb only suggest hydrocephalus.
- Proceeds to assess the *tone:* Hypotonia suggests cerebellar pathology while hypertonia suggests brain stem pathology.
- Then the *muscle power.*
- Then demonstrate the *posterior column sensations:*
  - Sense of position, sense of movements, sense of vibration
- Then perform *Romberg's sign,* if positive suggests sensory ataxia; while in cerebellar ataxia the child is off balance with eyes open and worse with eye closure.

Causes of Ataxia:

Acute	* Infection: virus e.g. chicken pox cerebellitis, bacteria e.g. brucellosis * Intoxication: drugs e.g. phenytion, antihistamines * Space occupying lesion, e.g. medulloblastoma, cerebellar tumour, brain stem tumour, neuroblastoma * Hemorrhage due to hypertension, AV malformation, blood disorders. * Trauma *Metabolic e.g. maple syrup urine disease (MSUD), hypoglycaemia * Seizure (epileptic ataxia) e.g. myoclonic encephalopathy
Chronic	* Static e.g. cerebral palsy, clumsy child syndrome * Progressive e.g. tumour, abscess, hydrocephalus, neurodegenerative disease
Intermittent	* Metabolic e.g. MSUD, Hartnup, multiple carboxylase deficiency * Vascular e.g. migraine, benign paroxysmal ataxia * Familial e.g. periodic ataxia (autosomal dominant) * Seizure * Conversion reaction

SUDDEN WEAKNESS OR INABILITY TO WALK (ACUTE PARALYSIS)

History:

- Verify the following features of the weakness:
  - Distal (neuropathic) or Proximal (myopathic)
  - Static or Progressive
  - Ascending (e.g. Guillain-Barre syndrome)
  - Diurnal variation (myasthenia gravis)
  - Bulbar symptoms (polio)
  - Sensory symptoms (neuropathic)
- Ask about preceding upper respiratory tract infection (URTI)
- Infectious disease, e.g. chicken pox, measles, infectious mononucleosis
- Back pain or muscle pain, which increases, by cough, sneezing, movement of spine (epidural abscess, disc prolapse, transverse myelitis).
- Symptoms of increased ICP, trauma to the back, or drug intake
- GIT symptoms, e.g. constipation with botulism or diarrhoea with poliomyelitis.
- Fever, e.g. epidural abscess
- Any change of voice, regurgitation, asymmetry of face, e.g. Guillain-Barre syndrome
- Vaccination, e.g. poliomyelitis, polio like illness.
- Any urinary or stool retention.
- Any history of ingestion of stored food (botulism)
- Going outside (desert) picnic e.g. tick paralysis.
- History of sickle cell anaemia, e.g. anterior spinal artery infarction

Clinical approach:

- The inability to walk could be due to true paralysis i.e. muscle weakness or due to pseudoparalysis secondary to pain. If it is due to true paralysis, it is either a part of quadriparesis (i.e. 4 limbs weakness) but more in the lower limb, which is either central or peripheral or paralysis of lower limbs only (paraplegia).

True paralysis:

A. Quadriparesis:
   1. Central causes e.g. encephalitits or hypoxic ischemic insult
   2. Peripheral causes:
      - Spinal cord e.g. poliomyelitis, polio like illness, asthmatic amyotrophy
      - Peripheral nerves e.g. Guillain-Barre syndrome.
      - Neuromuscular junction e.g. tick paralysis,
      - botulism, myasthenic crisis
      - Muscle e.g. periodic paralysis, myositis

B. Paraplegia:
   2. Central:
      - Para-sagittal meningioma, superior sagittal sinus thrombosis
   3. Spinal:
      - Compressive:
        - Infective e.g. epidural abscess, TB, osteomyelitis
        - Traumatic e.g. disc prolapse, discitis.
        - Tumour e.g. neuroblastoma
      - Non compressive:
        - Infective e.g. transverse myelitis
        - Vascular e.g. anterior spinal artery occlusion

Differentiating features of upper and lower motor neuron lesions

Feature	UMNL	LMNL
Site of the lesion	Cerebral hemispheres, cerebellum, Brain stem, Spinal cord	Anterior horn cell, Roots, Nerves, Neuromuscular junction, Muscles
Muscle weakness	Quadriplegia, Hemiplegia, Diplegia, Paraplegia	Proximal (myopathy) Distal (neuropathy)
Muscle tone	Spasticity / Rigidity	Hypotonia
Fasiculations	Absent	Present (particularly tongue)
Tendon reflexes	Hyperreflexia	Hypo / areflexia
Abdominal reflexes	Absent (depending on the involved spinal level)	Present
Sensory loss	Cortical sensations	Peripheral sensations
Electromyography (EMG)	Normal nerve conduction Decreased interference pattern and firing rate	Slow nerve conduction Large motor units Fasciculations and fibrillations

Pseudoparalysis: Inability to walk due to non neurological causes: e.g.

- Unrecognized trauma (fracture, contusion, sprain)
- Synovitis
- Osteomyelitis
- Arthritis

Clue:

- Normal muscle tone, reflexes
- Unilateral involvement, abnormal gait
- Focal tenderness can be commonly elicited

Differentiation between the most common causes of true paralysis:

Guillain-Barre syndrome	Poliomyelitis	Transverse myelitis
Symmetrical Flaccid all time Facial nerves involvement Sensory loss is variable Autonomic disturbance	Asymmetrical Flaccid all time Bulbar involvement No sensory loss No autonomic disturbance	Symmetrical Flaccid initially then spastic Normal cranial nerves Sensory level on trunk Usually normal

SUDDEN UNILATERAL WEAKNESS (ACUTE HEMIPLEGIA)

Clinical approach:

History of:

- URTI (tonsillitis)
- Skin rashes (purpuric, petechiae, maculo-papular)
- Contact with or encountering infectious disease
- Convulsion or loss of consciousness
- Trauma to head, neck or mouth
- GIT symptoms, e.g. diarrhoea with severe dehydration
- Cardiac symptoms (shortness of breath, cyanosis, tachypnoea)
- Symptoms of raised ICP
- Blood disorders, e.g. haemoglobinopathy

On examination:

- General:
  - Well or ill
  - Color (blue, pale, polycythaemic)
  - Skin rashes, neurocutaneous stigmata
  - Growth parameters: tall e.g. homocystinuria
- Neurological examination (stresses on)
  - Gait (ask the child to walk, then to run etc..) - hemiplegic gait
  - Assess the higher function (speech, cortical sensation etc..)
  - Cranial nerves, especially for facial asymmetry and hemianopia

- Motor system of upper and lower limbs for: asymmetry, tone, power, reflexes.
- Assess the sensation for sensory level

- Other systems: To find out the possible cause:
  - Examine the CVS as the commonest cause of acquired hemiplegia is vascular due to thrombosis, hemorrhage, embolism due to cyanotic congenital heart disease or due to hypertension.
  - Examine the ENT, e.g. tonsillitis or otitis media

Clue for levels :

- Cortical: Monoplegia ± speech defect (dysphasia) + loss of cortical sensation, + convulsion
- Subcortical: Monoplegia only
- Capsular: Monoplegia + homonymous hemianopia
- Brain stem: Mid brain: Contralateral hemiplegia + ipsilateral III, IV cranial nerve palsy
- Pons: Contralateral hemiplegia + ipsilateral VI nerve palsy
- Medulla: Contralateral hemiplegia + ipsilateral IX, X, XII nerve palsy
- Spinal cord:
  - Ipsilateral hemiplegia
  - Ipsilateral loss of deep sensation
  - Contralateral loss of superficial sensation with sensory level

Causes of sudden hemiplegia

1. Vascular: (stroke) sudden focal neurological deficit due to cerebrovascular accident:
   a) Infarction: Idiopathic
   b) Thrombosis:
      - Hyperviscosity: e.g. gastroenteritis with dehydration, diabetic ketoacidosis, polycythaemia
      - Hypercoagulability: e.g. sickle cell anaemia, idiopathic thrombocytopenia, post splenectomy, homocystinuria, arteriosclerosis, septicaemia, haemolytic uraemic syndrome, disseminated intravascular coagulation
   c) Embolism: Cardiac arrhythmia e.g. cyanotic congenital heart, mitral valve prolapse, myoxoma
   d) Spasm: Migraine, excessive swimming
   e) Vasculitis: e.g. systemic lupus erythematosus, Henoch-Schonlein purpura, Kawasaki disease, cervical infection (tonsillitis), Moyamoya disease
   f) Anoxic: e.g. status epilepticus, cardiac arrest, hypotension, airway obstruction
   g) Hemorrhage:
      - Idiopathic
      - AV malformation, rupture of aneurysm

- Hypertension
- Blood disease (ITP, coagulation diseases)
- Trauma, haemorrhage in tumour
- Chicken pox treated with steroid, pertussis

2. Infection: Encephalitis (herpetic), meningitis, abscess
3. Tumour
4. Trauma: accidental or non accidental

Causes of recurrent hemiplegia:

1. Sickle cell anaemia
2. Migraine
3. Mitral valve prolapse
4. Hemiparetic seizures
5. Moyamoya disease

FLOPPY INFANT SYNDROME

The infants who are floppy on handling. Clinically they present by delayed motor milestone i.e. delayed turning, setting, standing or because the mother feels he is floppy on handling or could be picked up during clinical assessment for other causes e.g. recurrent chest infection.

Types of Hypotonia

A. Central: is due to a lesion in the brain.

Clue:

- Child is not alert or not responsive, not interested to the surroundings.
- Normal spontaneous movements.
- Reflexes are normal or increased.
- Persistence of primitive reflexes.
- Fisting of hands, scissoring of legs on vertical suspension.
- Presence of organomegaly, e.g. congenital infection, storage diseases.
- Odd features of face (dysmorphic features).
- Presence of convulsion.

Causes:

1. Cerebral palsy - hypotonic or dystonic
2. Chromosomal aberration: e.g. Down's syndrome, Prader-Willi syndrome.
3. Cerebral malformation e.g. cerebellar hypoplasia
4. Benign congenital hypotonia (bottom shuffler)

B. Peripheral: Is due to a lesion in the spinal cord or peripheral nerves or neuromuscular junction or muscle.

Clue:

- Alert, responsive, interested to the surroundings
- Paucity or absence of spontaneous movements
- Reflexes are absent or diminished.
- Presence of muscle atrophy
- No abnormalities of other organs.

Causes:

1. Spinal cord: e.g. hypoxic ischaemic myelopathy, congenital tumour, spinal cord A-V malformation, injury (pre or intrapartum).
1. Anterior horn cell: e.g. spinal muscular atrophy, infantile neuronal degeneration.
2. Peripheral Nerves: e.g. congenital hypomyelinating neuropathy, giant axonal neuropathy, hereditary motor and sensory neuropathy.
3. Neuromuscular: e.g. myasthenia gravis, infantile botulism.
4. Muscle: e.g. congenital myopathy metabolic myopathy (Pompe's disease, carnitine deficiency, phosphorylase deficiency, cytochrome C oxidase deficiency) congenital muscular dystrophy, Warbrug syndrome, leucodystrophy.

C. ***Mixed:*** Due to a lesion of brain, with one component of the peripheral motor unit either muscle or nerve or anterior horn cell.

Causes:

Hypoxic ischaemic encephalomyelopathy, lipid storage disease, e.g. Tay-Sachs, Neimann-Pick, mitochondrial disease, peroxisomal disease, Pompe's disease, infantile neuroaxonal dystrophy, neonatal myotonic dystrophy, congenital muscular dystrophy, Lowe's syndrome, familial dysautonomia, Warburg (cerebello ocular dysplasia), methylmalonic aciduria, biotinidase deficiency, hypothyroidism

CLINICAL APPROACH:

History:

- Ask about fetal movements during pregnancy
- e.g. weak fetal movements (spinal muscular atrophy, congenital myotubular myopathy, carnitine deficiency)
- Polyhydramnios (spinal muscular atrophy, congenital myotonic dystrophy,congenital myotubular myopathy)
- Seizure (cerebral palsy, peroxisomal disorders, congenital muscular dystrophy with cerebral dysplasia, organic aciduria, amino acidopathy.
- Prenatal, perinatal, postnatal history to document anoxia, birth asphyxia
- trauma - mode of presentation, etc. ...
- Developmental milestones
- Course of hypotonia: progressive or regressive
- Hypotonia which improves with time:
    - Congenital myopathy
    - Prader-Willi syndrome
    - Cerebral malformation e.g. cerebellar hypotonia
- Family history of neuromuscular disease, death in neonatal or early infancy.

Examination:

- *Observe:*

- Posture of the child e.g. frog leg position

- Alertness, attentiveness of the child to the surrounding.
- Dysmorphic features of particular syndrome associated with hypotonia e.g. Down syndrome, Prader-Willi syndrome, Zellweger syndrome, congenital myotonic dystrophy
- Tongue for fasciculation (spinal muscular atropine), for size macroglossia (Pompe's disease, hypothyroidism, cytochrome C oxidase deficiency)
- Spontaneous movement:
- Observe the child for a while to see if he can move his limbs spontaneously or not, if the movement is more proximal or distal and if it affects upper or lower limb or both, then proceed to formal muscle power assessment.

• Maneuvers
  - Upper limbs: Shake test, then the range of movement at the joints, Scarf sign, and dorsiflexion of the wrist.
  - Lower limbs: Hip abduction >150 degree, toes-mouth sign, popliteal angle, ankle dorsiflexion.
  - Do the180 degree maneuver to assess the degree of hypotonia.
  - Examine the deep tendon reflexes: to differentiate between central and peripheral hypotonia

• Other systems: To find out a clue for possible cause of hypotonia
  Chest: e.g. bell shaped chest with paradoxical respiration (spinal muscular atrophy)
  CVS: e.g. cardiac lesions (Pompe's disease)
  Abdomen: e.g. organomegaly (congenital infections, Zellweger syndrome, storage disease), umbilical hernia (hypothyroidism), small genitalia (Prader- Willi syndrome)

CEREBRAL PALSY (CP)

Definition: Is a motor disorder resulting from a non-progressive insult to the developing brain during the perinatal or postnatal period (up to 2 years of life).

Causes: A variety of congenital and acquired disorders affecting the developing brain can cause cerebral palsy; however, up to 40% of cases are idiopathic.

A) Prenatal causes: (commonest after idiopathic)
- Cerebral malformation e.g. congenital cyst, fusion defect, abnormal migration of grey matter, congenital malformations.
- Congenital infection e.g. TORCHS
- Asphyxia
- Microcephaly
- Maternal phenylketonuria
- Fetal alcohol syndrome
- Prematurity

B) Natal causes:
- Asphyxia
- Birth injury

- Trauma
- Infection

C) Postnatal causes:

- Asphyxia
- Trauma (accidental or non accidental)
- Kernicterus
- Meningitis and encephalitis
- Status epilepticus
- Metabolic encephalopathy

Classification: Cerebral palsy can be classified according to the motor disorder (tone abnormality) or anatomical distribution and severity. Here, we shall mention the classification according to the American Academy for Cerebral Palsy.

1. According to motor disorder:
   - Spastic
   - Rigid
   - Hypotonic
   - Ataxic
   - Athetotic
   - Mixed
2. According to anatomical distribution:
   - Tetraplegia: Paralysis of the 4 limbs which are affected equally or sometimes upper limbs more than lower limbs
   - Paraplegia: Paralysis of lower limbs
   - Hemiplegia: Paralysis of one half of body where upper limbs are affected more than lower limbs
   - Diplegia: Paralysis of 4 limbs which affects lower limbs more than upper limbs
   - Triplegia: Paralysis of 3 limbs
   - Monoplegia: Paralysis of one limb
   - Double hemiplegia: Paralysis of both half of the body where one side is affected more than the other
3. According to degree of severity:
   - Mild
   - Moderate
   - Severe

   Severity is defined in terms of how disability affects the child's function, emphasizing on what the child can do rather than cannot do.

Clinical Presentation:

The clinical presentation depends on the pattern of cerebral palsy severity and the age of presentation to you. Early diagnosis could be easy in some cases while extremely difficult or even impossible in others. There are some symptoms and signs which leads you to suspect cerebral palsy

Symptoms:

- Arousal problems: Sleep disturbance, irritability, inconsolable crying, lethargy.

- Feeding problems: Sucking, swallowing difficulties (e.g. sucks the dummy but refuses feeds, takes a few sucks and spit them out, choking during feeds, regurgitation, vomiting, drooling).

Signs:

- Failure or delay to achieve normal motor milestones
    a. Manipulation like reaching, holding and transferring
    b. Gross motor like sitting, rolling, standing and walking
- Persistence of primitive neonatal reflexes after the expected time of disappearance.
- Abnormalities of posture and muscle tone. The mother might notice that the child feels stiff or floppy while handling or he has fisted hands after age of 2 months.
- It might be detected incidentally during routine screening or follow up for other problem like deafness.

N.B. Early recognition of cerebral palsy is important as deformity is always acquired. So it can be prevented or reduced.

Associated disorders:

a) Non motor problems:

- Behavior and psychological disorders e.g. sleep disturbance, irritability, poor concentration span, hyperactivity, recurrent screaming attacks.
- Visual problems: e.g.
    - Decreased visual acuity (30%)
    - Squint: It is a very common association which may be concomitant or paralytic (40%)
    - Optic atrophy with or without loss of vision (20%)
    - Error of refraction e.g. myopia
- Hearing impairment: it is mainly sensory neural deafness (10%)
- Mental retardation: IQ between 70 to 90 (60%)
- Epilepsy: This may be in the form of absence attack, drop attacks, grand mal or temporal lobe epilepsy. It depends on the type of cerebral palsy. Bilateral hemiplegic CP has the higher incidence of epilepsy (60%). While it is very rare or even absent in ataxic and dyskinetic CP.
- Educational problem: It depends on the type of cerebral palsy, the severity of cerebral palsy and IQ of the child. The cause of these educational disorders is due to speech defects, vision defect and hearing defect and/or mental retardation. They may need either physical handicapped or mental handicapped school.
- Speech disorder: Due to bulbar palsy with incoordination of tongue, palate and lips or mental retardation and/or hearing defects

- GIT problems:
    - Drooling due to bulbar palsy
    - Constipation: Immobilization and hypotonia
- Regurgitation and vomiting due to bulbar palsy with presence of gastro-oesophageal reflux or hiatus hernia.
- Recurrent chest infection due to frequent aspirations or immobilization which leads to hypostatic lung or scoliosis or chest deformity which interfere with ventilation.

b) **Motor problems:** Due to spasticity or rigidity with deformity

Management of cerebral palsy:

- **Investigations:** Cerebral palsy diagnosis depends mainly on careful history (prenatal, natal, postnatal, family history and developmental history) and thorough examination, which includes CNS, ENT, eye and musculoskeletal system. The investigations needed are guided by the findings on history and examination and should be directed to examine associated features and complications, as well as to establish the possible causes. e.g. audiometry (deafness), EEG (epilepsy), visual and auditory evoked response, TORCH, Brain CT or MRI, metabolic screen, chromosomal analysis.
- **Prevention:** From the etiology we can see most of the causes are acquired like birth asphyxia, birth trauma, haemorrhage, kernicterus and hyperviscosity which could be prevented if proper obstetric, perinatal, postnatal cares are given, regular follow up especially for at risk group.

- **Management:** It is multi-disciplinary approach - It is a team work management i.e. should be accomplished by cooperation of paediatrician, physiotherapist, psychologist, orthopedist, ENT, ophthalmologist, social worker. etc.

NEURODEGENERATIVE DISORDERS (NDD)

- Diverse and complex disorders with different classification systems.
- May have predominant white matter involvement (central and/or peripheral demylination) or gray matter involvement (neuronal loss or dysfunction).
- May have a mixed involvement particularly in the later stages of the disease, i.e. secondary demylination following progressive neuronal loss and secondary neuronal loss following progressive demylination.

Neurodegenerative Disorders	Inheritance patterns
1- Disorders predominantly involving the white matter	
- Canavan Disease	Autosomal recessive
- Alexander Disease	Sporadic
- Krabbe Leukodystrophy	Autosomal recessive
- Metachromatic Leukodystrophy	Autosomal recessive
- Pelizaeus Merzbacher Disease	X linked recessive
- Adrenoleukodystrophy	X linked recessive
- Multiple Sclerosis	Sporadic
2- Disorders predominantly involving the Gray matter	
- Menkes Kinky Hair Syndrome	X linked recessive
- Symptomatic Progressive Myoclonic Epilepsies (e.g. Unverricht-Lundborg disease, lafora disease)	Autosomal recessive
- Progressive Infantile Poliodystrophy	Autosomal recessive
- Sialidosis (Type I)	Autosomal recessive
- Neuronal Ceroid Lipofuscinosis	Autosomal recessive
- Mitochondrial Encephalopathies	Variable

NDD with preferential CNS involvement	Inheritance patterns
1- Disorders predominantly involving the basal ganglia	
- Juvenile Huntington disease	Autosomal dominant
- Dystonia Musculorum Deformans	Autosomal dominant
- Hallervorden Spatz Disease	Autosomal recessive
- Wilson Disease	Autosomal recessive
2- Spinocerebellar degeneration and related conditions	
- Friedreich Ataxia	Autosomal recessive
- Spinocerebellar Ataxia	Autosomal dominant
- Olivopontocerebellar Atrophy	Autosomal dominant
- Roussy Levy Disease	Autosomal recessive
3- Spastic paraplegia	
- Familial Spastic Paraplegia	Autosomal dominant
4- Peripheral neuropathy	
- Spinal Muscular Atrophy	Autosomal recessive
- Infantile Neuroaxonal Dystrophy	Autosomal recessive
- Charcot Marie Tooth Disease	Autosomal dominant
- Refsum Disease	Autosomal recessive

History taking

- Exclude perinatal complications (asphyxia, kernicterus, meningitis, head trauma)
- Family history of neurological disorders and early or unexplained deaths may indicate an undiagnosed inherited NDD.
- Diagnostic labels should not be taken for granted, as misdiagnosis is not uncommon, particularly true for cerebral palsy.
- Developmental history is needed to distinguish a static (non-progressive) from a progressive clinical course (loss of previously acquired milestones or regression).
- Other causes of developmental regression:
    - Behavioral syndromes (attention deficit hyperactivity disorder and autism)
    - Neuropsychiatric disorders (depression and child neglect)
    - Progressive visual impairment or hearing loss

- Intractable epilepsy (subclinical seizures)

Clinical examination

- Examination may be normal in the early stages.
- Abnormal behavior may be the only initial manifestation in the following disorders:
  - Metachromatic leukodystrophy
  - Juvenile adrenoleukodystrophy
  - Subacute sclerosing panencephalitis
- All eventually manifest clinical signs with careful follow up.
- The main differentiating features of predominantly white or gray matter NDD are summarized in the table.

Differentiating Features	White matter disorders	Gray matter disorders
Age of onset	Usually late (childhood)	Usually early (infancy)
Head size	May have megalencephaly	Usually microcephaly
Seizures	Late, rare	Early, severe
Cognitive functions	Initially normal	Progressive dementia
Peripheral Neuropathy	Early, demylination	Late, axonal loss
Spasticity	Early, severe	Later, progressive
Reflexes	Absent (neuropathy), or exaggerated (long tracts)	Normal or exaggerated
Cerebellar signs	Early, prominent	Late
Fundal examination	May show optic atrophy	Retinal degeneration
EEG	Diffuse delta slowing	Epileptiform discharges
Electromyography (EMG)	Slow nerve conduction	Usually normal
Evoked potentials (VEP, ABR)	Prolonged or absent	Usually normal
Electroretinograms (ERG)	Normal	Abnormal

- Detailed general and CNS examination is needed.
- Examination of eyes (the window of the brain) may give important diagnostic information as shown in the table.

Disorder or groups of disorders	Ocular abnormalities
Peroxisomal disorders	Optic atrophy
GM1, GM2, Niemann-Pick disease	Cherry red spot
Leukodystrophy	Optic atrophy
Mitochondrial disorders	Pigmentary retinal degeneration
Mucopolysaccharidosis	Corneal clouding
Mucolipidosis	Corneal clouding
Ataxia-telengiectasia	Conjunctival telangiectasia
Cockayne syndrome	Lenticular opacities (cataracts)
Wilson disease	Kayser-Fleisher corneal ring
Niemann-Pick disease (type C, D)	Vertical gaze palsy
Kearns-Sayre syndrome	Progressive external opthalmoplegia
Pelizaeus-Merzbacher disease	Pendular nystagmus

- Hepatomegaly and splenomegaly are evident in the neurovisceral sphingolipidosis, mucopolysaccharidosis, peroxisomal, and mitochondrial disorders.
- Cardiopathy occurs in mitochondrial disorders, friedreich ataxia, and mucopolysaccharidosis.
- Progressive renal failure occurs in fabry disease, sialidosis II, and Lowe syndrome.

Investigations

- Specific diagnosis is needed for treatment, prognosis, and genetic counseling.
- Investigations are directed towards identifying the underlying diagnosis and examining associated complications (e.g. seizures) as discussed previously.
- The findings on history and physical examination will guide the physician in selecting the required laboratory investigations.
- Metabolic acidosis on blood gas analysis could indicate organic acidopathies, urea cycle disorders, and mitochondrial encephalopathies.
- Skeletal survey may reveal specific bony abnormalities such as dysostosis multiplex in mucopolysaccharidosis.
- Neuroimaging, particularly brain MRI, is critical as it can show characteristic features and exclude malformations or slow growing brain tumors.
- Serum ammonia, lactate, pyruvate, amino acids, and urine for amino acids and organic acids would screen for most amino acid disorders, organic acidopathies, and urea cycle abnormalities.
- Selective use of specific diagnostic tests and enzyme assays is needed to reach a definitive diagnosis as summarized in the table.

Neurodegenerative Disorders	Diagnostic test
Canavan Disease	N-acetylaspartic acid (urine)
Alexander Disease	β-crystallin (CSF)
Krabbe Leukodystrophy	β-galactosidase (leukocytes / fibroblasts)
Metachromatic Leukodystrophy	Arylsulfatase A (leukocytes / fibroblasts)
Adrenoleukodystrophy	Very long chain fatty acids (VLCFA)
Mucopolysaccharidosis	Mucopolysaccharides (urine)
Mucolipidosis	Oligosaccharides (urine)
Menkes Kinky Hair Syndrome	Serum copper and ceruloplasmin
Lafora disease	Skin biopsy (intracytoplasmic lafora bodies)
Sialidosis (Type I)	α-neuraminidase (leukocytes / fibroblasts)
Neuronal Ceroid Lipofuscinosis	Skin, conjunctival, or rectal biopsy
Mitochondrial Encephalopathies	Lactate (CSF / blood), Muscle biopsy
Wilson Disease	Urine copper, serum copper and ceruloplasmin
Friedreich Ataxia	DNA studies (blood)
Spinal Muscular Atrophy	Muscle biopsy, DNA studies (blood)
Infantile Neuroaxonal Dystrophy	Nerve biopsy
Charcot Marie Tooth Disease	Nerve biopsy, DNA studies (blood)
Refsum Disease	Phytanic acid (blood)
Lesch-Nyhan disease	Hyperuricuria and hyperuricemia

Treatment

- Multidisciplinary team approach.
- Genetic counseling is important
- Treatable complications (epilepsy, feeding difficulties, gastro-esophageal reflux, spasticity, drooling, skeletal deformities, and recurrent chest infections).
- Specific treatments to counteract the offending metabolite, replace the dysfunctional enzyme, or vitamin therapy are summarized in the following table.

Neurodegenerative Disorders	**Specific treatment modality**
Krabbe Leukodystrophy	Bone marrow transplantation
Metachromatic Leukodystrophy	Bone marrow transplantation
Adrenoleukodystrophy	Glyceryl trioleate and trierucate, steroids for adrenal insufficiency, diet low in VLCFA, bone marrow transplantation
Mucopolysaccharidosis	Bone marrow transplantation, recombinant human α-L-iduronidase
Menkes Kinky Hair Syndrome	Copper sulfate
Mitochondrial Encephalopathies	Nicotinamide, riboflavin, dichloroacetate, L-carnitine, CoQ10
Wilson Disease	D-penicillamine, trietine, zinc acetate, liver transplantation
Refsum Disease	Reduction of phytanic acid intake
Lesch-Nyhan disease	Allopurinol

DEVELOPMENTAL EXAMINATION

Neurodevelopment is a continuous process by which one acquires learned skills. Although the development is dependent on the neurological maturation, the developmental examination is quite distinct from the neurological examination.

Developmental assessment is divided into four fields

- **Gross motor - locomotion.**
- **Fine motor - eye and hand control**
- **Language (receptive and expressive)**
- **Psychosocial**

Rules for developmental assessment

- Developmental assessment can be performed up to 6 years of age after which formal IQ testing should be done.
- Development follows an orderly progress. In a cephalo-caudal direction (head control, sitting, then standing etc.) e.g. child cannot sit, if there is no head control.
- Development of a particular skill requires loss of certain primitive reflexes, e.g. child can not sit if the tonic neck reflex persists.
- For the premature infant, the assessed developmental level should be adjusted in relation to chronological age during the first 2 years of life where a correction factor should be used.

During the first year - the correction factor is 100% i.e. subtract the number of the prematurity months from the chronological age (e.g. 8 months child who was 3 month premature should be scored on a developmental test as if he / she was 5 months (8 - 3 = 5 months).
During the second year - the correction factor is 50 % i.e. subtract half the number of prematurity months from the chronological age (e.g. 17 months child who was borne 2 months premature should be scored in developmental test as if he / she was 16 months.
i.e. $17 - \frac{2 \times 50}{100} = 16$ months

- During developmental examination:
    - One should be opportunistic to collect as much information as possible without disturbing the child.
    - One should have a plan rather than testing in haphazard manner.
    - One should be well familiar with the use of his tools i.e. cubes, toys picture, book etc.
    - One is advised to remember 1-2 milestones in all fields of development from different ages and be familiar with them.
- Always start by observation of what the child is doing before touching the child to do any maneuver, as it is unwise to handle the child with maneuver without getting more information from what he is doing.
- If the child is demonstrating a skill proceed to test for next skill i.e. if the child is walking, test for running and hoping.
- Primitive reflexes, should be checked until the child starts sitting and pivoting i.e. 8 - 11 months
- There is a range of age for the achievement of any developmental milestone. One should have the tendency to overestimate the age rather than to underestimate in order to avoid parental anxiety but of course it should be within the normal range.
- It is not always possible to assess the developmental age by one examination, especially when the child is sleepy, unwell, hungry or irritable due to any reason. In these circumstances the developmental assessment should by deferred to a later appropriate time.

Developmental reflexes
Are important reflexes that can be elicited in young infants as summarized in the table.

Developmental reflex	**Age of development**	**Age of disappearance**
Truncal incurvation	Birth	1-2 months
Rooting	Birth	3 months
Moro	Birth	5-6 months
Tonic neck	Birth	5-6 months
Palmar grasp	Birth	6 months
Adductor spread	Birth	7-8 months
Plantar grasp	Birth	9-10 months
Landau	5-10 months	24 months
Parachute	8-9 months	Persist

- Developmental reflexes can be abnormal when absent or weak (diffuse brain insults or drug effects). However, the reflexes that appears at birth are vigorous and complete only at term and may be weak or absent in the preterm (<37 weeks gestation) infant.
- If these reflexes persist or become exaggerated they may indicate upper motor neuron lesion. As well, asymmetry may be abnormal. For example, asymmetric moro reflex may indicate hemiplegia, brachial plexus injury, shoulder dislocation, or fractured humerous or clavicle.

ESSENTIAL MILESTONES

6 weeks – 3 months old

Gross motor	Head control (6 weeks - 3 months). Ventral suspension (head held in the level of the body briefly). Prone position: Pelvis flat and hips extended.
Fine motor and vision	Stares on looking at mother or examiner's face. Follows horizontally to 90°.
Language	Respond to rattle or bell 15 cm at ear level by quietening or turning to the sound. Startle response.
Social behavior	Smiles (6-10 weeks). Turns to regard the observer's face.

3 - 8 months old

Gross motor	Bears weight on legs while held upright (6 - 8 months) Downward parachute response (4 - 6 months) Sits with support (4 - 6 months) Sits without support (5 - 8 months) Can be pulled to sit (3½ - 6 months) Forward parachute response (7 - months) Crawls (6 - 9 months)
Fine motor and vision	Reaches out to grasp (palmar) (3 - 6 months) Transfers and mouths (5 - 8 months) Fixes on small objects (5 - 8 months) Follows falling toys (6 - 8 months)
Language	Vocalizes (4 - 6 months) Polysyllabic babbling (6 - 10 months) Responds to whisper 1 foot away at ear level (4 - 8 months) Distraction hearing test
Social behavior and play	Puts everything to mouth (4 - 8 months) Hand and foot regard (4 - 8 months) Plays peek-a-boo (6 - 10 months)

12 months old

Gross motor	Pulls to standing position (6 - 10 months) Walks holding furniture (7 - 13 months) Walks alone (10 - 15 months)
Fine motor and vision	Points with index finger Casts (throws) (9 - 15 months) Pincer grasp (9 - 14 months) Holds 2 bricks and bangs them together (7 - 13 months)
Language	Turns to sound (10 - 12 months) Uses ?mama? and ?dada? (11 - 20 months, 50% children by 15 months) Understands several words Distraction hearing test
Social behavior and play	Drinks from cup (10 - 16 months) Indicates wants (10 - 15 months) Plays pat-a-cake (8 - 13 months) Waves good-bye (8 - 13 months)

1 – 2 years old

Gross motor	Walks backwards (12 - 22 months) Carries toys while walking. Climbs stairs with 2 feet per step holding the rail (14 - 22 months) Climbs onto chair
Fine motor and vision	Delicate pincer grasp (10-18 months) Scribbles (12-24 months) Turns pages Builds tower of 3 or 4 bricks (16-24 months)
Language	Jabbers continually Utters 3 or more words other than mama and dada (10 - 21 months) Points to eyes, nose and mouth (14 - 23 months) Obeys simple instructions ù ?close the door? (15 months - 2½ years)
Social behavior and play	Holds spoon: gets food to mouth (14 months - 2½ years) Explores environment (13 - 20 months) Takes off shoes and socks (13 – 20 months) Indicates toilet needs

2 – 3 years old

Gross motor	Climbs stairs unaided with one foot per step and descends with 2 feet per step Jumps in place (21 months - 3 years) Kicks ball (15 - 24 months)
Fine motor vision and vision	Picks up hundreds and thousands Imitates vertical line (18 months - 2½ years 9) Builds tower of 8 bricks (2 months - 3½ years)
Language	Uses plurals (20 months - 3½ years) Gives name Hearing test; speech discrimination test
Social behavior and play	Plays alone Eats with spoon and fork Puts on clothes Dry throughout day

3 – 4 years old

Gross motor	Runs fast Climbs stairs in adult manner Paddles tricycle (2 - 3 years) Stands on one foot for 1 second (2 -3 years)
Fine motor and vision	Copies circle (2 ½ years - 3 years) Threads beads well Matches 2 colours
Language	Uses prepositions (3 years - 4½ years) Uses sentences of 4 words Gives full name, sex and age (2½ - 4 years)
Social behavior and play	Eats with knife and fork Goes to toilet alone Dresses with supervision (2½ - 3½ years) Washes and dries hands (2 - 3½ years) Separates from mother easily (2 - 4 years)

4 years old

Gross motor	Hops on one foot for 2 meters (3 - 5 years) Climbs ladder, tree, slide Stands on one foot for 5 seconds (3 - 4½ years) Walks heel-to-toe (3½ - 5 years)
Fine motor and vision	Copies cross and square Imitates bridge of 3 bricks Draw man with 3 parts
Language	Speaks grammatically (3 - 4 years) Counts up to 10 Gives full name, age and address Recognizes colors (3 - 4½ years)
Social behavior and play	Shares toys Brushes teeth Dresses without supervision (3½ - 5½ years)

5 years old

Gross motor	Walks downstairs (one foot per step) Bounces and catches ball (3½ - 5½ years) Walks backwards heel-to-toe (4 - 6 years)
Fine motor and vision	Copies triangle (5½ - 6 years) Draws a man with all feature (4½ - 6 years) Copies 3 steps with 6 bricks
Language	Speaks fluently and clearly Hearing test (Audiometry)
Social behavior and play	Comforts friends in distress Chooses own friends Dramatic group play

- **Screening test:** Hand/eye coordination and cognitive development could be tested by drawing and copying abilities. The following are common shapes and the appropriate ages of their development:

Scribble	1 1/2 - 2 1/2 years	
Circle	2 1/2 - 3 years	
Cross	3 1/2 - 4 years	
Square	4 1/2 - 5 years	
Triangle	5 1/2 - 6 years	
Rectangle	6 years	
Diamond	7 years	

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MUSCUOSKELETAL SYSTEM

Musculoskeletal pain in children could be of inflammatory or non-inflammatory origin. Non-inflammatory causes of pain are much more common than those of rheumatic origin. Although they are often in sequential in the final diagnosis, their identification and differentiation from rheumatic, infectious, malignant and degenerative disease is essential for appropriate therapy.

For evaluating a child with musculoskeletal pain, history and physical examination are the foundations of diagnostic approach, and the laboratory investigations provide limited help.

KEY POINTS IN HISTORY:

The history should be able to answer the following questions:

1-what is the character of the pain:

A. How long the pain been present,
B. What makes it worse, and what makes it better,
C. Dose the pain interfere with function? (Rheumatic fever arthritis pain usually very painful)
D. Is their diurnal variation in the severity of the pain? (Morning stiffness in juvenile rheumatoid arthritis)
E. Is the pain presents at night? (Growing pain typically at night)
F. Quality of pain (sharp, aching, deep, etc)
G. Dose the painful part looks different?
H. Constant or migrating? (Rheumatic fever)

2- History of associated symptoms:

A- Fever: (*intermittent fever e.g. systemic onset JRA, nocturnal fever e.g. tuberculosis or malignancy, continuous or remittent fever e.g. Kawasaki disease, undulant fever e.g. brucella)*
B- Weight loss (suggestive of malignancy, systemic illness as IBD, SLE)
C- Depression (e.g. SLE)
D- Abdominal pain (IBD, JDM, HSP, SLE).
E- Diarrhea (suggestive of reactive arthritis, chronic shigellosis, salmonellosis, compylobacter, or IBD)
F- Muscle weakness (dermatomyositis)
G- Skin rash (look for rashes and joints at the end of this chapter)
H- History of proceeding upper respiratory tract infection (suggestive of rheumatic fever)

3- Is there family history of musculoskeletal disease?

4- What are the social, emotional, and educational circumstances?

KEY POINTS IN PHYSICAL EXAMINATION:

Golden rules:

1-The child's modesty and dignity should be preserved all times
2-The child's permission to be examined should be sought informally
3- Never hurt the child, and he or she should be reassured that examination would be stopped if it causes pain.
4- Never forces a joint during examination.

The initial examination should be able to help you to answer the following questions:

A- Is the pain localized to soft tissue (? Trauma), bone (? trauma, malignancy, or osteomyelitis) or joint (arthritis)?
B- Is there more than one painful site?
C- Is there tenderness as well as pain? (Painful but not tender area suggests the possibility of referred pain
D- Is the appearance of the affected are abnormal (e.g. Swelling, erythema, and heat suggest inflammation)

General physical examination is an essential part of the evaluation of any child with musculoskeletal pain, since pain is frequently a manifestation of systemic disease. Follow the following format of steps for clinical examination:

INSPECTION

General:

- Fever
- Pallor (anemia, malignancy)
- Skin rash
- Nail pitting (psoriasis)
- Lymphadenopathy
- Hepatosplenomegaly (JCA, malignancy, sickle cell anemia)
- Physical growth (growth failure is seen in most chronic inflammatory diseases of the musculosckeletal system and some times the side effect of medical therapy)
- Muscle weakness
- Eye involvement (JCA)

Local (it should be done at rest and during movement):

- Number and distribution of the joint involvement (symmetry)
- Joint swelling (fluid within a joint causes loss of normal contour i.e. bony landmark of the joint)
- Skin changes (Erythema implies associated periarticular inflammation)
- Deformity or contracture
- Muscle wasting
- Sinus formation, cautery marks

PALPATION:

Before touching the child you should explain to him/her what you are going *to do and examination will be discontinued if pain persists*

Feel the joint and periarticular area for:

- Warmth
- Tenderness (joint-line/capsular tenderness signifies arthropathy, periarticular tenderness signifies enthesitis or bursitis)
- Effusion
- Synovial thickening (boggy swelling)
- Crepitus (fine crepitus is due to inflammation of the tendon sheath, bursa or synovium, coarse crepitus reflects cartilage or bone damage)

RANGE OF MOVEMENTS:

Start by active movement then proceed to passive movement (synovitis produces usually similar reduction of both movements, passive movement far greater than active movement suggests a muscle/tendon/motor problem)

The range of movement or angle should be assessed from the neutral position (use your own joints as reference)

Detailed individual joint examination will follow.

ASSESSMENT OF FUNCTIONAL ABILITY:

Functional assessment for each joint region is assessed by observation during normal usage and activities of daily living

Don't forget to assess the gait

KNEE JOINT EXAMINATION

INSPECTION:

- Comment on posture of the limb, inflammation of the synovium will cause the child to adopt the joint position of maximum intracapsular capacity (minimum tension), usually semiflexion
- Signs of inflammation (redness, shiny skin)
- Joint swelling or deformity
- Cautery marks or sinus formation
- Muscle wasting of the quadriceps
- Observe the standing posture, and the gait

PALPATION:

- **Skin temperature:**- The temperature of swollen joints should be compared by touch with that of the other limb
  The back of the hand is a sensitive thermometer for comparing skin temperature above, over, and below the inflamed joint
- **Tenderness: -** Precise localization of tenderness is the most useful sign in determining the cause of the patient problem:
  Joint line tenderness: signifies arthropathy
  Periarticular point tenderness away from the joint signifies bursitis or enthesopathy
- **Effusion: -**
  - **Mild effusion:** This can be detected by the bulge sign. Keep the knee straight in extension, any fluid in the antero-medial compartment of the knee is massaged up into the suprapatellar pouch, and normal depression medial to the patellar tendon is seen to bulge as the fluid accumulates there
  - **Moderate to large effusion:** This is detected by the patellar tap. Keep the knee straight, and then the suprapatellar pouch is emptied by pressure with one hand while the other hand (index and middle fingers) taps the patella against the underlying femur by a sharp downward pressure.
  - N.B. large effusion can be differentiated from synovial thickening by squeezing the knee swelling while palpating on each side of the patella with the other hand. In case of effusion, transmission of the fluid impulse from hand to hand is felt.

RANGE OF MOVEMENTS:

Notice any limitation of movements (normal range from 0^{O} to 180^{O} of flexion. In females few degree of hyperextension is possible)
Palpate for any crepitus during joint movement

TESTS OF STABILITY:

Not to be performed if the knee joint is acutely inflamed.

A- *Cruciate ligaments*:
- Keep the knee flexed at 90° and maintain this position
- With both hands free, check the hamstring muscles are relaxed
- To test anterior cruciate the tibia is grasped just below the knee and is drown forwards (pull)
- To test posterior cruciate this movement is reversed (push)

B- *Collateral ligaments:*
- Keep knee in full extension
- The patient's ankle is held between the examiner's elbow and side leaving both hands free to abduct and adduct the tibia on the femur while keeping the knee straight.
- Normally no lateral movements occur but patient feels localized pain
- When ligament is lax click is heard when pressure is released

C- *The McMurray Test:*
- The object of this is to test the stability of the semilunar cartilage. This will induce a torn cartilage to engage between the tibia and the femur by reproducing the mechanisms, which originally caused the displacement
- When this happens the patient experiences the typical; symptoms and palpable (occasionally audible) "click" or "cluck" results.
- Let the patient be relaxed
- To examine the right knee the examiner stands on the right side of the couch
- The knee is then flexed to limit the patient will tolerate.
- While pressing on the outer side of the knee with the left hand the knee is extended while tibia is alternately, internally and externally rotated by the right hand

MEASUREMENTS:

Look for discrepancy between the two sides:
- A tape measure record of muscle wasting can be done by circumfrential measure at point (6-10 cm) above the superior patellar pole on both sides
- Measure the maximum calf circumference
- Measure leg length (from anterior superior iliac spine to medial malleolus)

GENU VARUM (BOW LEGS)

- Seen in 1-3 year old age group
- Examination reveals diffuse bowing of the lower extremities with an increased distance between the knees, which is accentuated on standing

Causes of bow legs:

- Physiological
- Rickets
- Blount's disease
- Epiphysial dysplasia

GENU VALGUM (KNOCK KNEES)

- Wile the standing child is noted to have increased distance between the feet while the medial aspects of the knees touch one another
- It is more common in females
- In patients with genu valgum (knock-knees) the distance between the medial malleoli when the child stands with feet parallel and knees just touching, can be measured (intermalleolar distance)
- It is pathological if it is more than 5 cm in child over 8 years

Causes of knock-knees:

- Physiological
- Rickets
- Ligamentous laxity
- Renal osteodystrophy

HIP JOINT EXAMINATION

INSPECTION:

- Undress the patient to underpants and examined walking, standing and lying.
- Inflamed painful hip tends to be held in slight flexion, abduction and external rotation.

PALPATION:

The hip joint is deeply seated and joint swelling is not usually apparent.

- Palpate the joint landmarks for tenderness, and warmth.
- Localized tenderness over the anterior part of the hip may be due to joint inflammation or bursitis.
- Localized tenderness over the lateral aspect of the greater trochanter could be due to bursitis

RANGE OF MOVEMENTS:

Except for hip extension all movements are best examined while the patient lying supine.

- Flexion (approximate 120^{O}) is tested with knee flexed
- Abduction (approximately 45^{O}) is frequently lost in hip disease
- Adduction: (approximately 30^{O}) is best assessed by crossing one leg over the other.

- Internal rotation: (approximately 45^{O}) is one of the earliest and most reliable signs of hip disease. It is best examined while both knee and hip are flexed at 90 degree.
- External rotation: (approximately 45^{O})
- Extension: check it while patient in prone position. While you immobilize the pelvis and the lumbar spine by one hand use the other hand to extend the hip.
- Any pain on resisted movements in association with localized pain and tenderness indicates tendenitis. For example pain on resisted adduction is typical of adductor tendentious and pain on resisted abduction is typical of gluteal tendenitis

Thomas' test: indicates a fixed flexion deformity of the hip:

- Flex the good hip fully, observing with the other hand that the lumbar spine is flattened.
- Lifting the thigh on the affected side with flexion of the knee indicates a positive test.

TESTS OF STABILITY:

Trendelenburg's Sign:

- This sign demonstrates that the hip abductors are not functioning
- It is useful in the late stages of congenital disease of hip when the patient is walking and in assessing the disability in polymyositis involving the lower limbs
- When normal subjects stand on one leg the glutei contracts so that the opposite side of the pelvis is tilted up slightly.
- If patient stands on the affected leg (the action of glutei are deficient) the opposite side of the pelvis will tilt downward and balance can be maintained only by leaning over the side of the lesion.

CONGENITAL HIP DISLOCATION (CDH)

Hip should be examined at birth, 6 weeks, 6-8 months and 18 months

Risk groups for CDH:

- Commoner in girls
- Commoner in first babies
- Family history of CDH
- Children with congenital foot deformity
- Breech presentation
- Cesarean section
- Oligohydramnios
- Fetal growth retardation

- All children with the above history should be examined carefully for CDH
- Test for CDH (*Ortolani and Barlow's maneuver*) look for details in the neonatal Chapter.
- In child with **unilateral dislocation** look for the following signs:
  - Leg posture: the thighs tend to be held in partial external rotation, flexion and abduction

- Limb shortening: above knee shortening on the affected side
- Asymmetry of the thighs: skin creases may be asymmetrical when checked in supine and prone position (not very reliable)
- Flattening of the buttock: may appear on the affected side in prone position
- Limitation of abduction: persistent and less than 75 degree (the most important sign)
- In child with **bilateral dislocation:**
- The signs as described, although there is no normal hip for comparison
- A perineal gap may be present.

Gait:

- 20% of children with CDH will not walk at 18 months of age
- 80% will walk and stand at normal age
- Child with unilateral CDH will limp or fall towards the affected side
- After 2 years the child can not balance on the affected leg
- In bilateral CDH the gait is waddling.

LEG LENGTH DISCREPANCY

Measurement of leg-length:

- Apparent shortening is measured between fixed point (xiphoid process or umbilicus) and the tip of the medial malleolus.
- True shortening is measured from the anterior superior iliac spine to medial malleolus.

Causes of discrepancy:

1- Hemihypertrophy:
   - Wilm's tumor
   - Beckwith Wiedemann syndrome
   - Neurofibromatosis
   - Diastematomelia
   - Klippel-Trenaunay-Weber syndrome
   - Silver-Russell syndrome
   - Local causes (cavernous hemangioma)
   - Idiopathic

2- Juvenile chronic arthritis
3- Skeletal Dysplasia
4- Perth's disease
5- Slipped capital femoral epiphysis
6- Radiation therapy
7- Lymphatic system obstruction (e.g., elephantiasis)

ANKLE JOINT AND FOOT EXAMINATION

- **The ankle joint**
  - The ankle joint is hinge joint
  - The ankle joint should have normal dorsiflexion and plantar flexion of about 30 °

- **Subtalar Joint:**
  - In the inversion and eversion of the hind foot the movements take place mainly in the subtalar joint
  - Her the examiner stabilizes the distal leg with one hand and grasp the heel with other hand to move the foot into inversion and eversion
  - Normal range is between 20-30 °

- **Midtarsal joint:**
  - The inversion and eversion of the fore foot take place in the mid-tarsal joint, between the talus and calcaneous posteriorily and the fore foot anteriorly
  - Her the heel is held in one hand and the fore foot is held in the other, then the fore foot is adducted and abducted in relation to the hind foot.
  - Normal range is 30-40 °

FLAT FOOT (PES PLANUS)

- Flat foot is a foot configuration characterized by an increased planter contact area
- It is best examined while the child is in standing posture and look from behind, lateral and front.
- It is usually asymptomatic, but some times associated with pain on weight bearing usually localized to the medial side of the arch.
- The pronated flat foot is often accompanied by mild genu valgum, slight flexion at the knees and increased lordosis. As a result of these mechanical strains, pain in the ankles, knees, or lower back might occur
- In adolescents, the Achilles tendon may shorten, thereby limiting ankle dorsiflexion.

PES CAVUS

It is a complex deformity composed of:

- High arched foot
- Clawing toes
- Inversion of the fore foot
- Varus of the hind foot

It is often a manifestation of neurological disease

Causes:

- Idiopathic 10%
- Neurologic 90%:
  - Spinal muscular atrophy

- Muscular dystrophy
- Cerebral palsy
- Frederick's ataxia
- Poliomyelitis

To detect the deformity, while the child is standing look for the foot from behind, lateral and front
Do pressure print method: Ask the child to stand on powdered glass.

HYPERMOBILITY OF THE JOINTS

- The term benign hypermobility is used to describe children in whom generalized hypermobility is associated with musculoskeletal pain
- It represents an extreme variation of normal range of joint motion
- It is not associated with underlying connective tissue disease like Marfan's or Ehler's Danlos syndrome

Criteria for establishing the diagnosis of hypermobility (Carter-Wilkinson criteria):

1. Opposition of thumb to flexor aspect of forearm
2. Hyperextension of fingers parallel to extensor aspect of forearm
3. Hyperextension of elbows or knees by more than 10 degree
4. Excessive dorsiflexion of ankle and eversion of foot

Diagnosis require presence of at least three criteria

TALIPES EQUINOVARUS

This is typical clubfoot deformity. It is characterized by three primary components:

1. The entire foot is positioned in planter flexion (*equinus*)
2. The hind foot is maintained in a position of fixed inversion (*varus*)
3. The forefoot exhibits an adductus deformity of tendon combined with supination

The typical idiopathic congenital clubfoot must be differentiated from similar deformity secondary to:

- Spinal cord tethering
- Myelodysplasia
- Degenerative neurological conditions.

TORTICOLLIS

It is tilting of the head due to muscle spasm

- Ask the child whether it is painful or not.
- Ask him if possible to do full range of neck movements

Then examine:

- Eyes for squint
- Mouth for pharyngitis, tonsillitis, and retropharyngeal abscess
- Face for asymmetry
- Pallor
- Neck for swelling, tenderness
- Shoulder for Sprengel's deformity, Klippel-Feil syndrome
- Assess the gait (ataxic gait suggestive of brain tumor)

Important causes:

- Sterno-mastoid tumor
- Trauma (sublaxation of atlanto-axial joint), do cervical spine –x-ray
- Posterior fosse tumor (not very uncommon)
- Eye ocular torticollis (superior oblique ocular muscle paralysis)
- Soft tissue: - cervical adenitis, retropharyngeal abscess
- Rheumatic "stiff neck" or wary neck (myositis of viral origin-local tenderness)
- Sandifer's syndrome: - reflux oesophagitis episodes associated with movements of the neck (torticollis) in patients with cerebral palsy look for pallor (iron deficiency anemia due to chronic blood loss).
- Drugs (e.g., metechlopromide) which causes extrapyramidal manifestations).

TOE WALKING

Bilateral

1. Normal variation up to 3 years of age
2. Cerebral diplegia
3. Duchen muscular dystrophy
4. Spinal muscular atrophy
5. Autism

Unilateral

1. Unilateral congenital hip dislocation.
2. Spastic hemiplegia.
3. Congenital shortening of tendo achilles.

EXAMINATION OF THE SPINE FOR SCOLIOSIS

- Let the child stand erect with feet together.
- Check symmetry of the shoulders.
- Ask the child to bend forward at the waist 90^{O}, let the child touch his or her toes.
- The examiner sitting behind the child will see a hump in case of fixed scoliosis

Causes of scoliosis:

1- Postural (scoliosis disappearing when the child bends over), 80%
   - Idiopathic
   - Unequal leg length
   - Unilateral muscle spasm

2- Structural (scoliosis persisting when the child bends over), 20%
   - Bone: hemivertebrae, Sprengel's deformity
   - Ligament: Marfan's syndrome
   - Muscle-muscular dystrophy
   - Neurogenic: spina bifida, poliomyelitis, Frederick's ataxia, and cerebral palsy
   - Post irradiation

PAIN AMPLIFICATION SYNDROMES IN CHILDHOOD

1) GROWING PAINS:

Age at onset: 4-to 12 years
Sex ratio: Equal
Symptoms: Deep aching, cramping pains in thigh or calf; usually in evening or during nigh, and often interrupt sleep; never associated with limb and symptoms disappear by morning; bilateral; responds to massage and analgesia
Signs: Physical examination is normal
Investigations: Laboratory examination normal

2) PRIMARY FIBROMYALGIA SYNDROME:

Age at onset: adolescence to adulthood
Sex ration: Girls > boys
Symptoms: Generalized fatigue, diffuse ill-defined musculoskeletal aching and stiffness, anxiety, depression, disturbed sleep patterns
Signs: Tender points at characteristic sites (see the figure 2)
Investigations: laboratory examination normal

Tender point sites according to the American College of Rheumatology 1990 criteria for the classification of fibromyalgia,

- *Occiput* (2) - at the suboccipital muscle insertions.
- *Low cervical* (2) - at the anterior aspects of the intertransverse spaces at C5-C7.
- *Trapezius* (2) - at the midpoint of the upper border.
- *Supraspinatus* (2) - at origins, above the scapula spine near the medial border.
- *Second rib* (2) - upper lateral to the second costochondral junction.
- *Lateral epicondyle* (2) - 2 cm distal to the epicondyles.
- *Gluteal* (2) - in upper outer quadrants of buttocks in anterior fold of muscle.
- *Greater trochanter* (2) - posterior to the trochanteric prominence.
- *Knee* (2) - at the medial fat pad proximal to the joint line.

Pain in 11 of 18 tender point sites on digital palpation, is needed for the diagnosis.

3) REFLEX SYMPATHETIC DYSTROPHY:

Age at onset: Late childhood and adolescence to adulthood
Sex ratio: Girls > boys
Symptoms: exquisite superficial and deep pain in the distal part of extremity, exacerbated by passive or active movement
Signs: Diffuse swelling, tenderness, coolness, and mottling; bizarre posturing of the affected part
Investigations: osteoporosis, bone scintigraphy abnormalities, laboratory examination normal.

EXAMPLES OF PAIN INTENSITY RATING SCALE

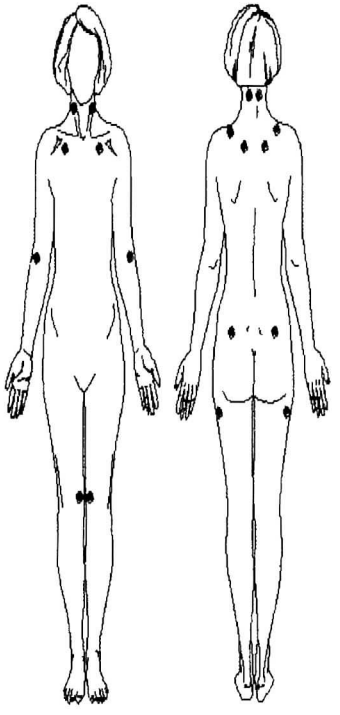
[Pain Scale]

Place a straight; up and down mark on this line to show how much pain you have

No pain	Little pain	Medium pain	Large pain	Worst possible pain

Figure 1: Pain assessment scale in children, Word graphic rating scale from Tesler, Savedra, Holzemer and Wilkie (1991).

Figure 2: tender point sites in fibromyalgia



SKIN RASHES AND THE JOINTS

Skin lesions	Causes
*Erythema marginatum	- Rheumatic fever
*Erythema nodosum	- Inflammatory bowel disease, TB etc
*Erythema chronicum migrans	- Lyme disease
*Erythema infectiosum (slapped face)	- Parvovirus
*Maculopapular	- Rubella, EBV, Adeno, hepatitis B
*Vesicles over the joint	- Gonococcemia
*Vasculitis	- HSP, Kawasaki disease, SLE etc
*Purpura and bruising	- Acute leukemia, hemophilia etc
*Evanescent maculopapular rash "Salmon Like"	-Rheumatoid rash in systemic onset JCA
*Butterfly malar rash	- SLE
*Heliotrope rash over the metacarpo-Phalangeal joints	- Dermatomyositis
*Scaly erythematous plaques	- Psoriasis

QUICK ASSESSMENT OF CHILD WITH JUVENILE CHRONIC ARTHRITIS

GENERAL EXAMINATION:

- Look for pallor, rash, lymphadenopathy, hepatosplenomegaly
- Temperature (look for signs of infection)
- Joint deformity
- Muscle wasting

JOINTS:

QUICK CLINICAL EXAMINATION CAN BE DONE AS FOLLOWS:

ASK THE PATIENT TO DO:

1. *Flex the neck to each side*: looking for pain and restriction of movements
2. *Open the jaw wide and move it from side to side*. Palpate tympanomandibular joint while opening his or her mouth and feel for click. Look for micrognathia
3. *Place both hands behind the head with elbows back*. External rotation and abduction are the earliest most severely affected glenohumeral movements.
4. *Place both hands out in front, palms down, fingers straight, and with elbows at 90 ^{O} at the sid*e. Inspect for abnormalities particularly swelling, deformity, attitude, and skin changes at the distal radioulnar joint, wrists, MCP's and interphalangeal (PIP's) joints.
5. *Turns the hands over (supination)*. Testing proximal and distal radioulnar joints. Inspect the palmar aspects for wasting, skin changes, flexor tenosynovitis swelling
6. *Make a tight fist with each hand*. Observe ability to curl fingers tightly into palms.
7. *Place the tip of each finger onto the tip of the thumb in turn*. Observe for fine precision pinch.
8. *Squeeze across the 2nd to 5th metacarpals.* Look for tenderness while patient lying supine on bed,
9. *Passively flex the hip and knee while holding the knee.* To test for hip flexion

10. *Passively internally and externally rotates the hip*. This is a sensitive test for hip disease
11. *Press downs the patella and palpate for effusion in each knee*
12. *Squeeze all metatarsals* to test MTP joints

GAIT:

- Any limping
- Any leg length discrepancy due to chronic monoarthritis (especially knee)

GROWTH:

- Check growth centiles (short stature as an effect of the disease or chronic use of steroid)

EYES:

Needs very careful assessment by ophthalmologist especially in case of pauci JCA:

- Look for irregular pupil, cataract, and band keratopathy
- Iridocyclitis
- Retinopathy (if patient using chloroquine)

SIGNS OF STEROID TOXICITY:

- Hypertension
- Growth retardation
- Features of Cushing's syndrome
- Osteoporosis
- Proximal myopathy
- Cataract
- Diabetes mellitus
- Gastric irritation
- Protein wasting, edema, metabolic alkalosis
- Susceptibility to infection
- Psychosis

CAUSES OF MONOARTHRITIS:

- **Acute monoarthritis:**
  - Septic arthritis
  - Oligoarticular JCA
  - Reactive arthritis
  - Leukemia
  - Neuroblastoma
  - Hemophilia

- **Chronic monoarthritis**:
  - Oligoarticular JCA
  - Juvenile ankylosing spondylitis
  - Juvenile psoriatic arthritis
  - Villonodular synovitis
  - Sarcoidosis

CAUSES OF POLYARTHRITIS:

- Polyarticular and systemic JCA
- Juvenile psoriatic arthritis
- SLE
- Reactive arthritis
- Rheumatic fever (migratory)
- Arthritis of inflammatory bowel disease.

REFERENCES:

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Radiological Imaging

Sensible selection of imaging investigations is of a major importance.
A non scientific approach is when selection is based upon trials and errors, ie, the examination request aims at the hope that something will support or refute, or something will turn up.
The right approach is that selection should be based upon.

a- Clinical presentation and laboratory data.
b- Awareness of radiological modality being requested, this is achieved by good communication between clinicians and radiologists.

This would result in carrying out the appropriate imaging technique and renders the radiologist's interpretation meaningful.

Main radiological Modalities:

1- Conventional x-ray:
   - Plain radiography.
   - Barium contrast (or alternative contrast) radiography.
   - Urography.
   - Cholangiography.
   - Angiography.
2- Ultrasound.
3- Computerized tomography Scanning.
4- Magnetic Resonance Imaging.
5- Radionuclide Imaging.

Conventional Radiography

- The principle of conventional radiology is based upon the differential absorption of different body tissues to beams of x-ray, which is a type of ionizing radiation that hinders significant biological effects.
- If beams pass through air (least absorbed tissue) → blackening of corresponding areas in the film.
- While if they pass through calcified ie, calcium containing structures (most absorbed tissues) → whitening of corresponding areas in the film...

In between densities are primarily fat and soft tissues, therefore limited visualization for four distinct densities: Gas, soft tissues, fat and calcified structures.

Advantages:

- Most accessible and practical modality.
- Non costly.
- A primary tool for baseline assessments and follow up evaluations.

Disadvantages:

- Use of ioizing radiation.
- Whole thickness two dimensional, ie, structures is seen overlapping.
- Limited density differentiation.

Indications:

(A) Plain radiograph : efficient primary imaging modality for:
- Chest diseases.
- Bones e.g. fractures and bone diseases.
- Abdominal conditions e.g. acute abdomen, calcifications.

(B) Urography :

- Appearance and function of urinary tract.

(C) Barium (or alternative contrasts) studies: swallow, meal, follow through and enema: for opacification and evaluation of esophagus, stomach, small and large bowel respectively.

(D) Cholangiography: opacification of biliary system.

(E) Angiography: vascular contrast opacification.

Ultrasound

Use of very high frequency sound transmitted to body tissues by a transducer, echoes reflected form tissue interfaces are picked up by the same transducer, and converted to electrical signal visualized on a television monitor.

Main indications:

- Imaging of superficial structures e.g. thyroid and small parts.
- Determination whether a structure is solid or cystic.
- Evaluation of solid organs lesions.
- Quantitative and qualitative evaluation of blood flow in vessels and vascular lesions.

Advantages:

- Lack of ionizing radiation.
- Real-time nature of examination.
- Multiplanar imaging capabilities, with great flexibility in selection of imaging planes and ease of altering these planes.
- Non costly.

Disadvantages:

-Limited densities:

-Operator dependency of acquiring the images.

Computed Tomography

- Provides detailed anatomic information valuable in diagnosis and disease management. CT is a technique that mathematically constructs a digital cross-sectional image by assimilating tissue absorption information obtained from multiple trans-axial x-ray projections. The data is thence converted to an image for display by computerized processes.
- Large number of different densities are discriminated by CT scanning, ie, high contrast resolution.
- Majority of studies require administration of intravenous contrast media for better characterization of lesions and delineation of anatomical structures. Common exceptions include hemorrhagic lesions and renal stones.

Advantages:

- High contrast resolution, ie, discrimination of subtle differences in densities.
- Good spatial resolution, ie, appreciation of small structures.
- Very fast scanning is acquired, three dimensional images can be reconstructed, and reformatted images at different planes (multiplanar acquisition) can be obtained.

Disadvantages:

- Relatively costly when compared with conventional x-ray and ultrasound.

- Use of ionizing radiation.
- Need of I.V. contrast material makes it limited in cases of renal impairment with increased creatinine levels and also in contrast allergy and atopies.

Indications:

(1) Excellent modality for all systems imaging (CNS, musculoskeletal, chest, abdomen and pelvis, whenever information acquired by simpler techniques is not sufficient.
(2) Evaluation of vascular structures and related diseases "CT angiography".

Preparation:

- Explanation of nature and physical requirement of the examination, e.g. necessity of immobility and cooperation.
- Patients under age of 4 years usually need proper sedation and I.V canula insertion.
- If I.V contrast is to be administered, then fasting (preferably for 4-6 hours) is advised to avoid aspiration.

Magnetic Resonance Imaging MRI

- An exquisitely sophisticated imaging modality with a very high contrast resolution, i.e, discriminates between subtle contrast differences.
- Basic principle is based upon the alignment of nuclei (protons) of certain elements of the body (mainly hydrogen atoms) in the direction of a strong external magnetic field. Subsequently, a radiofrequency (RF) pulse is a applied with a specific resonating frequency to result in a proportion of (protons) changing their alignment and flip through a specific preset angle, then they relax to their initial alignment after recovery from the (RF) pulse. The process of relaxation after flipping - in terms of alignment and arrangement, ie, phasicity- is responsible for induction of a radio signal which builds up an image.
- Image is created depending basically upon two relaxation times namely T1 and T2; however there are many other sequences with a bewildering variety of, names and acronyms.

Advantages:

(1) High contrast resolution and spatial resolution renders its diagnostic value outstanding.
(2) Multiplanar, with direct acquisition in any chosen plane.
(3) No ionizing radiation and no adverse biologic effect from diagnostic MRI have been reported.

Disadvantages:

- Relatively expensive and lengthy modality.
- A lot of body devices e.g., pacemakers, clips or intra ocular metallic foreign bodies are MR noncompatible which results in variable contraindications.
- Image degradation by unavoidable movements from breathing, cardiac pulsation and peristalsis.

MRI is rapidly developing technology with evolving techniques to speed up scan times and to limit effects of motion by use of various gating devices. Cardiac gating is already widely available.

Indications:

Detailed imaging of:

- Brain.
- Spinal cord.
- Bones and Joints: although calcified tissues do not generate any signal at MRI, MRI produces images of bone marrow and soft tissues.
- Pelvic organs.
- Biliary system.
- MRCP → magnetic retrograde cholangiopancreaticograply.
- Urinary tract.
- Cardiac Imaging.
- Vascular structures.
- MR arteriography (MRA).
- MR venography (MRV).

Selected Studies in Gastrointestinal Tract Imaging

Barium Studies:

Use of contrast media (e.g. barium which is an inert opaque material) to opacify lumen of a desired portion of the alimentary tract.

It is either performed as:

(1) A single contrast study; to evaluate the caliber and outline as well as filling defects at the area of interest.

(2) Double contrast study; to evaluate mucosal pattern for distorted mucosal pattern or ulcerations e.g. Crohn's disease.

Risks

- Suspension of perforation is a major contraindication, as escape of barium into peritoneal cavity or pleural cavity is a life threatening condition and may result in severe hypovolemic shock with around 50% mortality despite treatment.
- Treatment consists of I.V fluids, steroids and antibiotics.
- Out of 50% survivals → 30% develop peritoneal adhesions and granulomata.

Therefore, it is imperative that in such conditions where there is a risk of perforation, a water soluble contrast media is the optimal resort, e.g. gastrografin or an equivalent in instances like.

- Suspected perforation.
- Meconium illus.

On the other hand, however, water soluble contrast media are contra indicated in : **Suspected aspiration**: As there is a risk of developing pulmonary edema.

- In such cases non-ionic low osmolar weight contrast media (LOCM) is the contrast of choice.
- Another risk of water soluble contrast media is **hypovolemic shock** particularly in children due to their high osmolarity, therefore, resort to (LOCM).

Diagnostic procedures;

Contrast studies : Like contrast swallow, meal , follow through and enema for evaluation of esophagus, stomach, small intestines and large intestines respectively to evaluate conditions that are congenital or aquired.e.g. tracheoesophageal fistula, duodenal atresia, stenotic segments, meconium ilius, malrotation, and Hirschsprung disease .

Therapeutic procedures.

(A) Enema reduction of intussusceptions → a procedure which should only be attempted in full consultation with the surgeon in charge of the case.
(B) Therapeutic water soluble contrast enema in meconium ilius.

SELECTED STUDIES IN GENITOURINARY TRACT IMAGING

(1) Excretion Urography: "Intravenous Pyelography"

- Practical examination for evaluation of function and anatomy of urinary tract. An intravenous contrast material is introduced and gradual and sequential secretion by glomerular filtration into kidneys, followed by excretion into pelvicalyceal system, ureters and then urinary bladder is assessed by sequential imaging.
- Preferably : LOCM " low osmolar contrast media " are used particularly in high risk groups which are →
    - Infants and young children.
    - Renal or cardiac failure patients.
    - Patients with diabetes or sickle cell disease.
    - History of previous allergic reactions or atopies.
- Preparation:
    - No food at least for 5 hours.
    - Proper premeditation for allergic patients with steroids.

(2) Micturating Cystourethrogram (MCUG):

- Study of urinary bladder contrast filling with urethral pacification during micturation done by infusion of contrast material slowly through a catheter inserted into urinary bladder via urethra.

Indications:

- Vesicoureteric reflux.
- To study and evaluate congenital and aquired abnormalities of the bladder and urethra.

(3) Nuclear Urinary Tract Studies:

I. Static renal Scintigraphy :

→ Functional evaluation by use of 99 TC-2.3 dimercaptosuccinic acids (DMSA) or 99TC-glucoheptonate.

Indications:

- Assessment of renal function.
- Demonstration of ectopic renal tissues.
- Assess reflux nephropathy.
- Demonstrate congenital anomalies and mass lesions.

II. Dynamic renal scintigraphy:

- Use of 99 Tc-MAG-3.
- Or 99TC-DTPA.
- Or Hippuran (123 I complex).

Indications:

- Assessment of obstructive versus non obstructive dilatation of collecting system.
- Diagnosis of acute tubular necrosis.
- Differentiates hydronephrosis from multicystic dysplasia.
- Detects renal artery stenosis.

Image Interpretations

Approach to a Chest X-ray.

The entire film should be evaluated and not only the chest.
CBA approach is therefore suggested to ensure complicity of evaluation.
Where; A: Abdomen.
B: Bony cage, with soft tissue evaluation.
C: Chest.

Chest: Check:

(1) Trachea and mediastinum.

- Cardiac shadow usually lies in the middle with two thirds to the left and one third to the right (variable degrees do exist).
- Centralization of mediastinum: (Mediastinal shift) either; away from the lesion e.g.→ Pneumothorax, hydrothorax, diaphragmatic hernia. Or towards the lesion e.g. massive pulmonary collapse, lung fibrosis.
- Shape of cardiac shadow and position of aortic arch.

(2) Lung Fields :

- Symmetric translucency of comparable zones (upper, middle and lower zones) of the lungs bilaterally.
- No abnormal opacities e.g. consolidation, infiltrates, collapse.etc.
- Costophrenic angles being sharply clear and translucent.
- Normally visualized bronchovascular markings which should be most prominent at hilum, less prominent in middle third and almost absents in lateral third.

(3) Diaphragm :

- Normally: curved upwards medially.
- Rt. Hemidiaphragm is usually higher than left because the heart creates pressure element upon left lung and not because of the liver displacing right lung upwards.

(4) Bony cage (and soft tissues):

- Ribs, clavicles and vertebral column: check for lytic lesions (bone resorption) or sclerotic lesions (bone formation) or fractures.
- Evaluate soft tissues for any abnormalities e.g soft tissue emphysema or swelling.
- Crowdening or over separation of ribs by assessing intercostal spaces.
- Clavicles: absence, defects.
- Kyphosis/ scoliosis of vertebral column.

Abdomen:

- Check-gas pattern? ilius or obstruction , free air.
- Abnormal calcifications.
- Situs inversus.

***Approach to plain abdominal film*:**

The entire film should be evaluated ABC approach is suggested for complicity of interpretation.

A- Abdomen
B- Bone.
C- Chest – visualized part.

Abdomen:

Check

- Normal air distribution in bowel or extra luminal gas (free gas).
- Abnormal calcifications e.g. renal stones or adrenal clacifications.

- Soft tissues (liver, spleen, Kidneys, stomach... etc).
- Psoas muscles shadows → if obscured, think of retroperitoneal process occurring.

Bones:

Check for:

- Fractures in bony compartment.
- Bone deformities e.g. scoliosis.
- Bone defects or sclerosis.

Chest:

- Usually lower chest is visualized check consolidation, , free air under diaphragm → upright film.

Approach to skull x-ray:

Basically, lateral and frontal skull vies check for:

A- Intact bones: check for any fractures (linear lucent lines) or bone defects e.g. metastasis or histiocytosis.

B- Shape of bones: size is better assessed clinically.

1- Frontal Bossing: seen in hydrocephalus and thalassemia.

2- Craniosynostosis (craniostenosis).

- Acrocephaly (Oxycephaly): towering of head due to premature exposure of metobic suture.
- Brachycephaly: short head in anteroposterior diameter due to premature closure of sagittal suture.

CT scan studies interpretation

- Depending on window setting at console monitor, the structures of interest will stand out more clearly e.g. mediastinal window, lung window, bone window, and soft tissue window.
- Density (attenuation) is measured by hounsfield units (HU).

Brain CT scan:

- Normality of structures should be appreciated by proper knowledge of normal appearance and normal anatomy.
- Fresh blood e.g. subdural, epidural, subarachnoid, or intracerebral hemorrhage appears hyper dense i.e., high attenuation than brain tissue.
- After 2 weeks → fades to isodense ie, similar density to brain tissue.
- After 4 weeks → hypo dense, i.e., lower in deasity than brain tissue.
- Intravenous administration: required for visualization of pathological processes. e.g. neoplastic or inflammatory lesions with contrast crossing blood brain barrier causing their enhancement. Also required for evaluation of vascular structures and vascular lesions.
- The lesions appear either by density differences or their mass effect upon adjacent structures e.g. effacement of sulky sub cortical lesion or displacement of ventricles.

Chest CT scan:

- **Mediastinal window :**
  - With I.V contrast required for delineation of vascular structures and any vascular anomalies.
  - Lymph nodes and soft tissue lesions are better appreciated.
- **Lung window :**

- Lung parenchyma architecture should be maintained.
- Better visualization of focal lesions e.g. consolidations, atelactasis, calcifications and mass lesions. e.g. tumors.
- Better visualization of diffuse lesions e.g. interstitial lung diseases and military shadowing, particularly with high resolution CT.

Abdomen and Pelvic CT scan:

- Awareness of morphology and sizes of abdomen and pelvic structures is mandatory.
- Lesions appear in solid organs as :
  (a) Diffuse involvement e.g. or density of the liver in hemosiderosis and fatty infiltration respectively.
  (b) Focal density alteration with and without mass effect. e.g. tumor or infarction respectively.

This is appreciated by use of intravenous contrast material.
Stomach and bowel loops should be a pacified by oral water soluble contrast medium to differentiate between these structures and other pathological processes.

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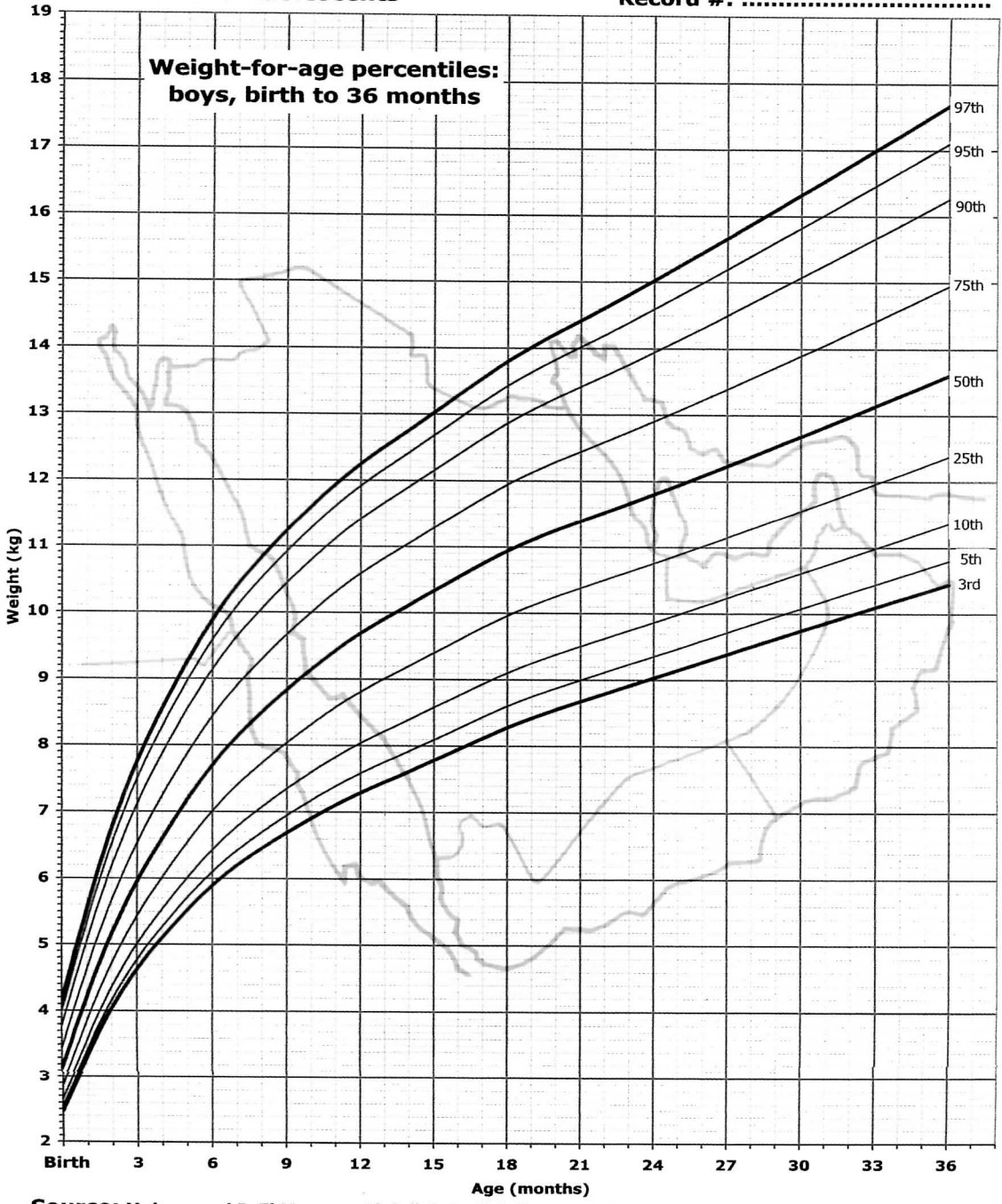
Appendix 1

Growth Chart

The Growth Charts for Saudi Children and Adolescents

Name:.....

Record #: .....



Source: Mohammad I. El Mouzan, Abdullah A. Al Salloum, Abdullah S. Al Herbish, Mansour M. Qurashi, Ahmad A. Al Omar. Health Profile for Saudi Children and Adolescents (No. AR-20-63). King Abdulaziz City for Science and Technology 2007, Riyadh, KSA.

NB: The age is based on Gregorian calender.

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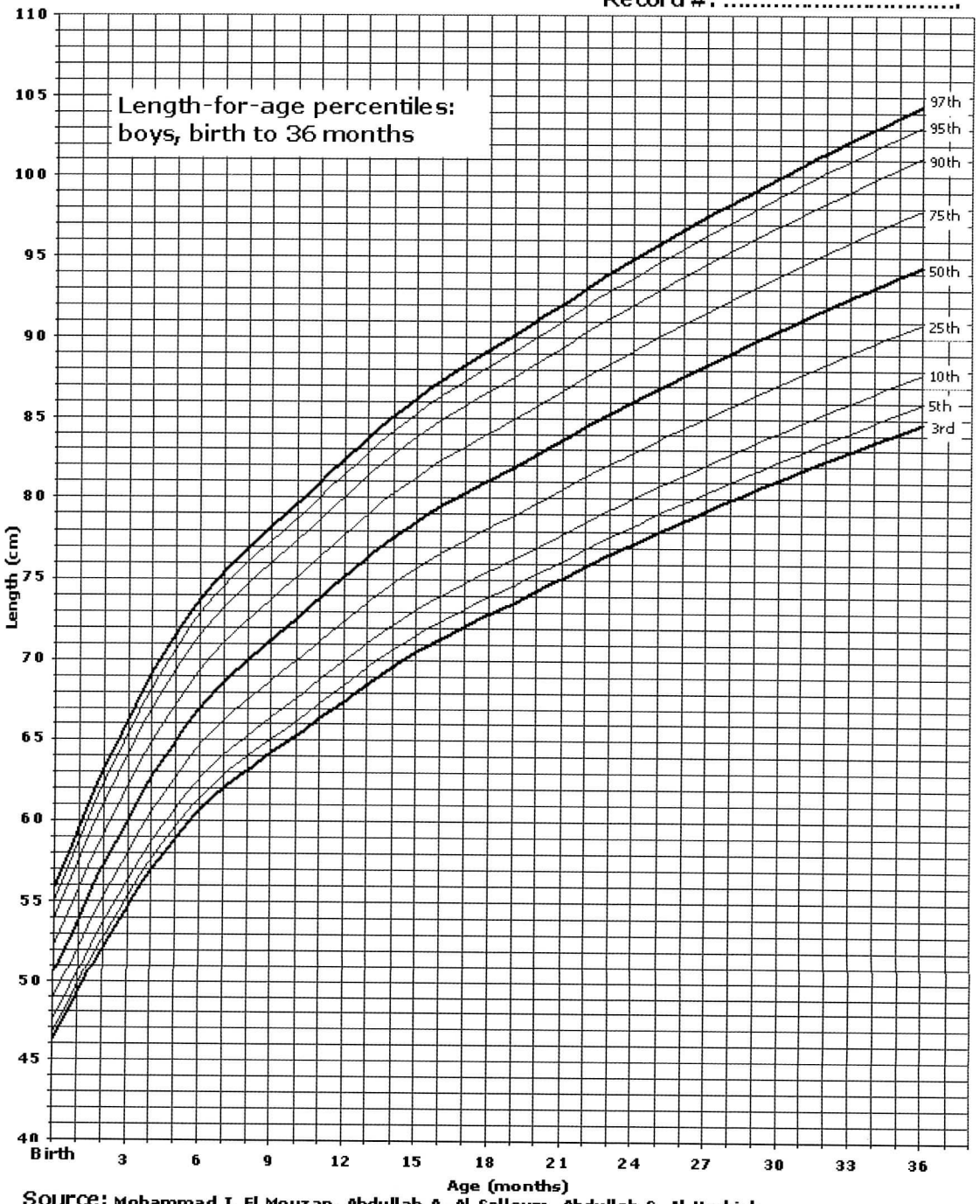

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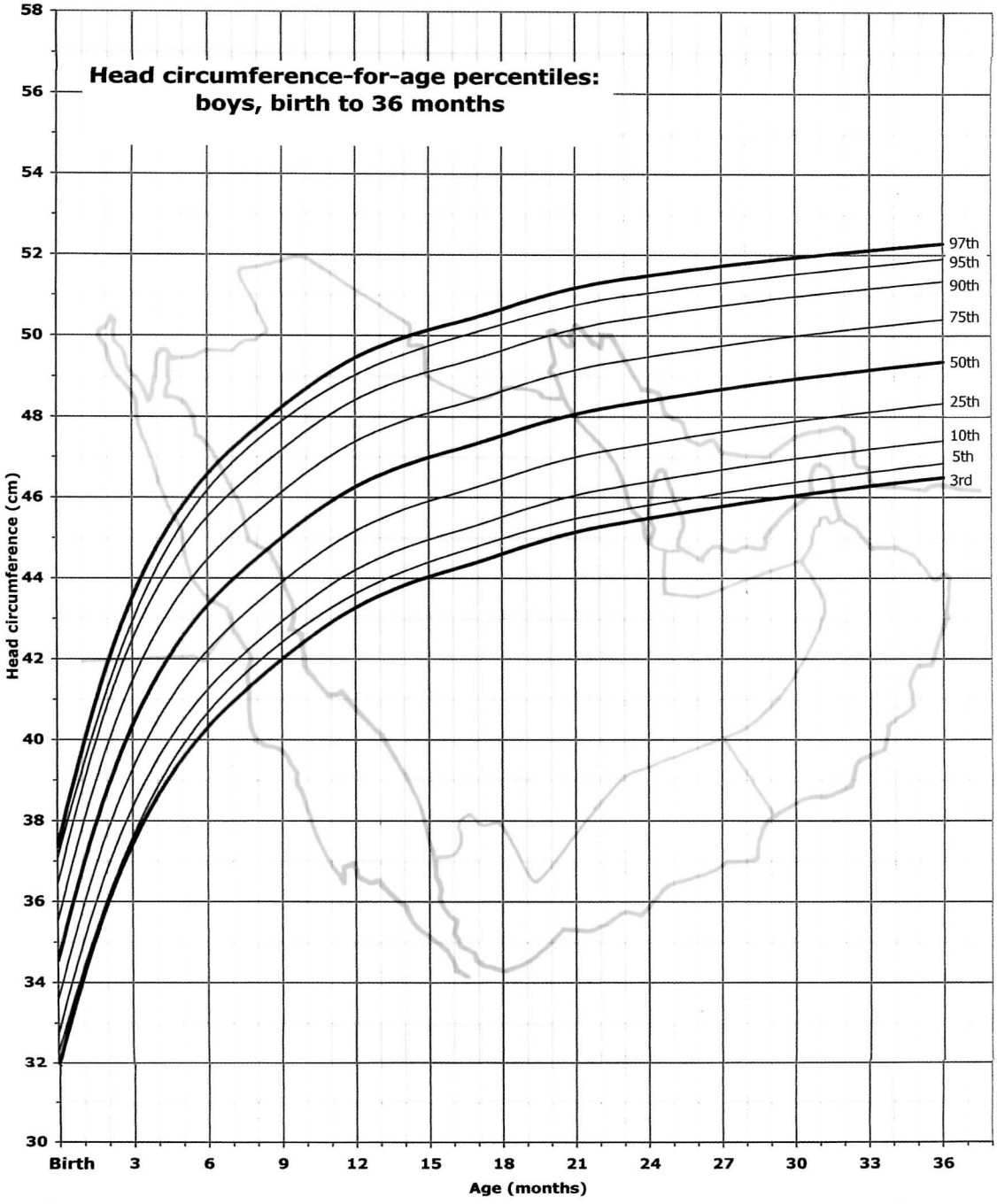

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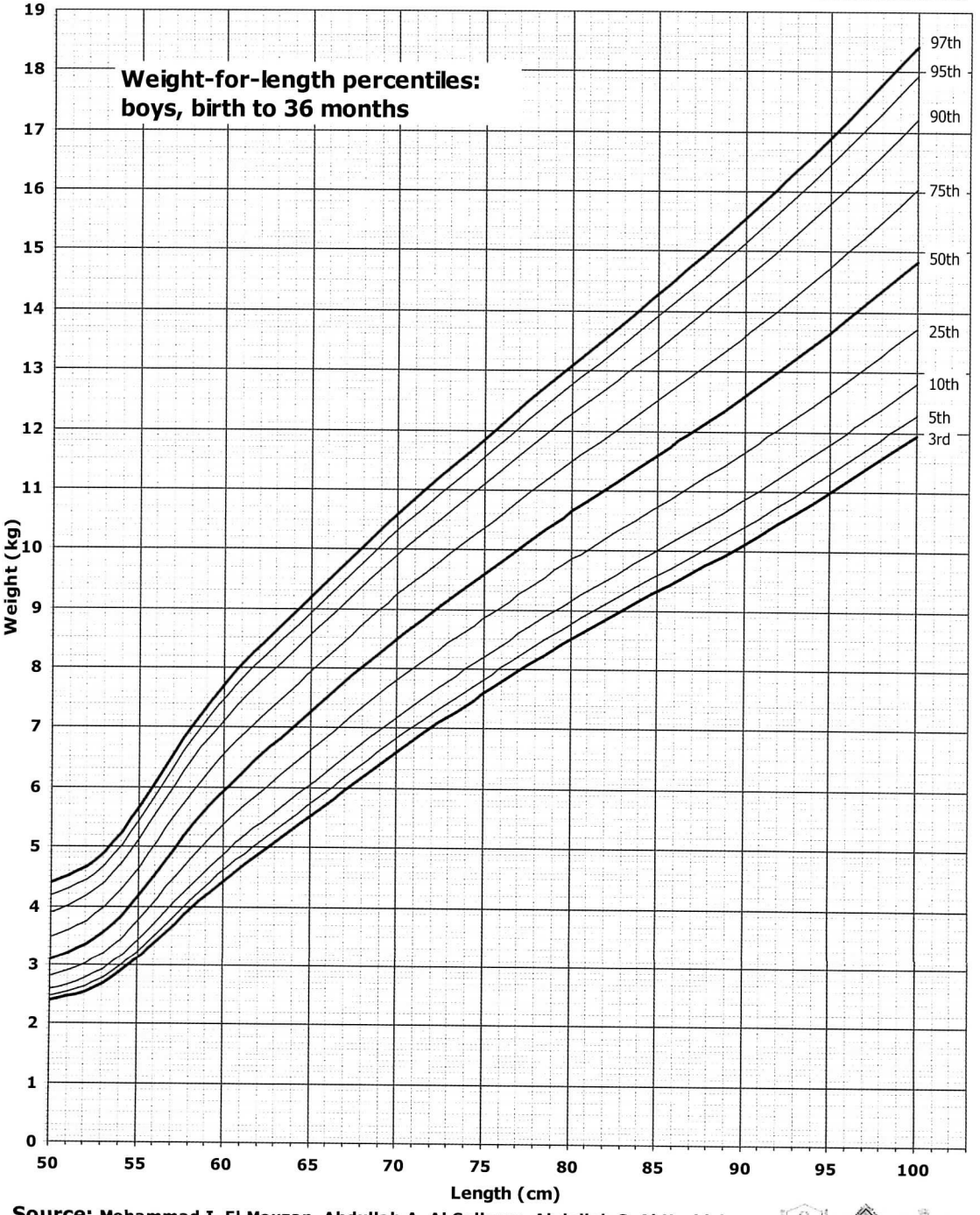

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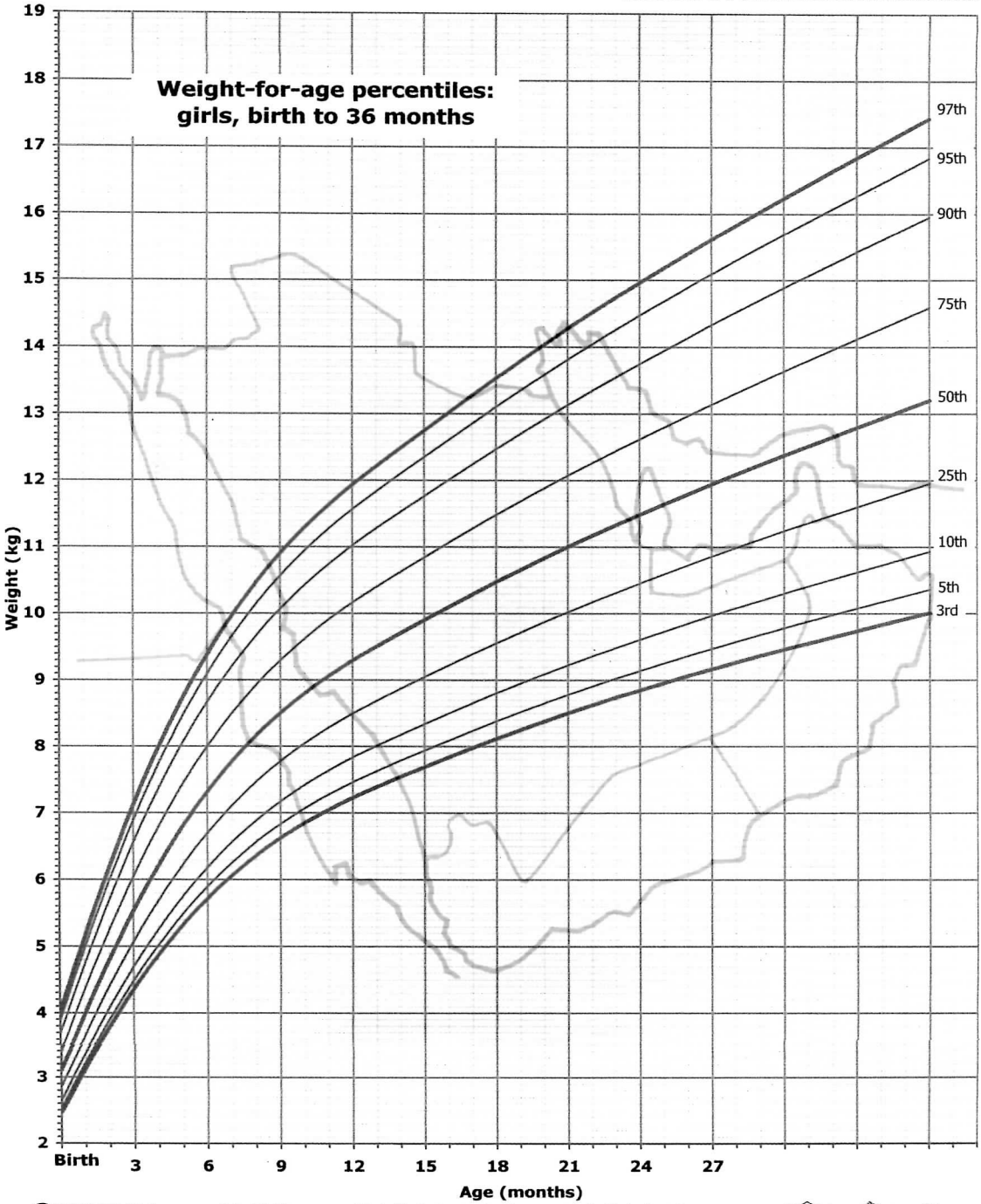

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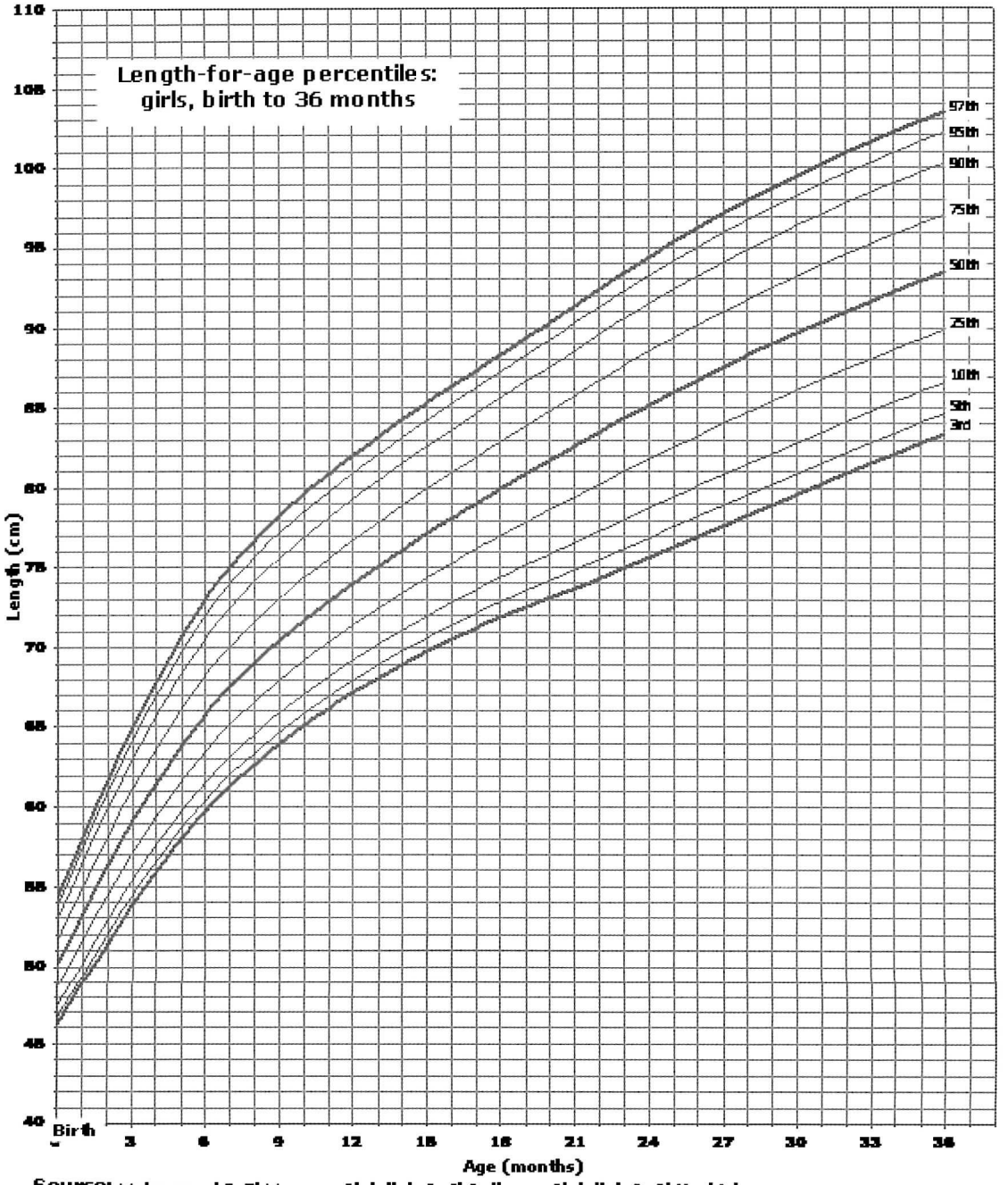

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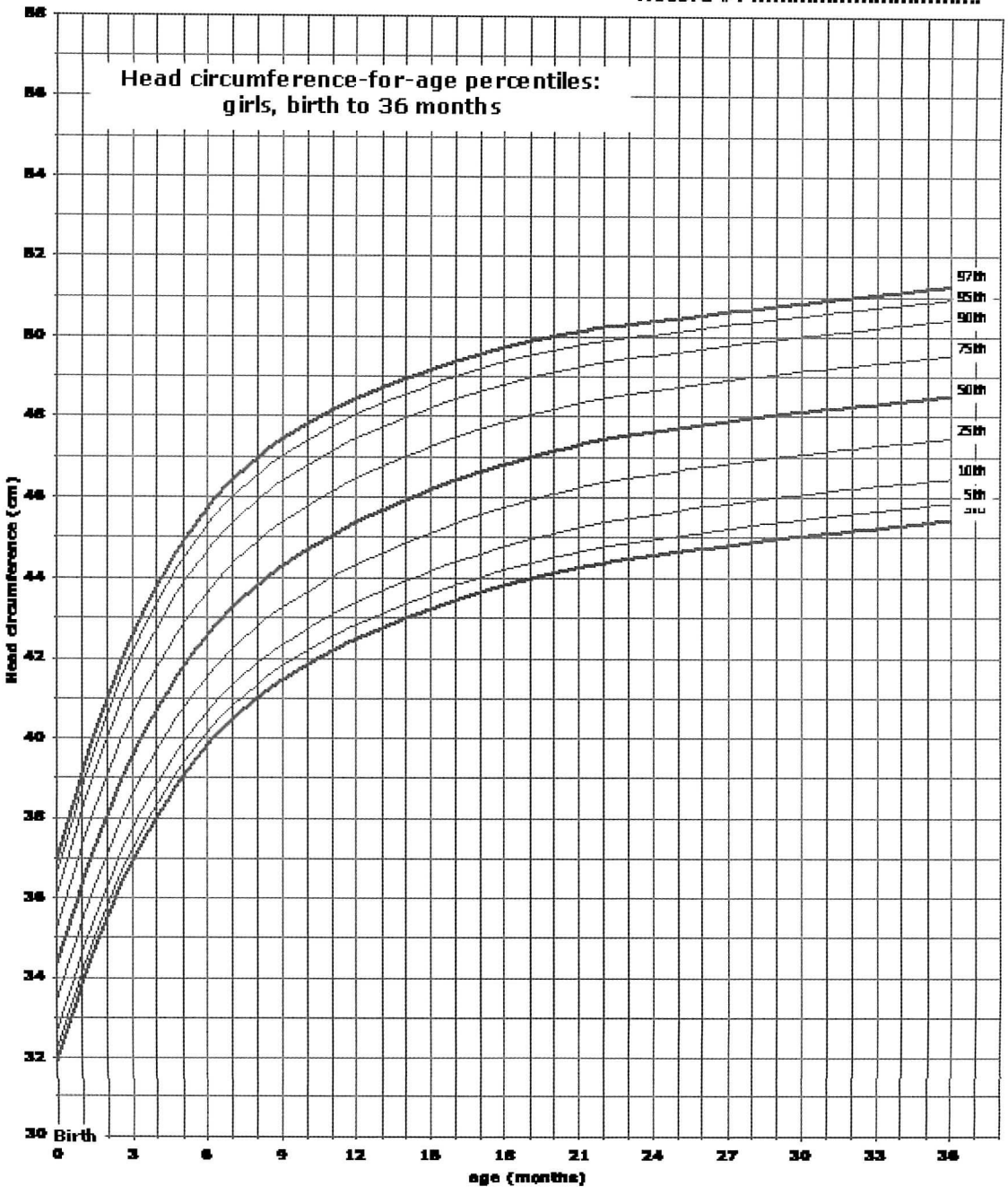

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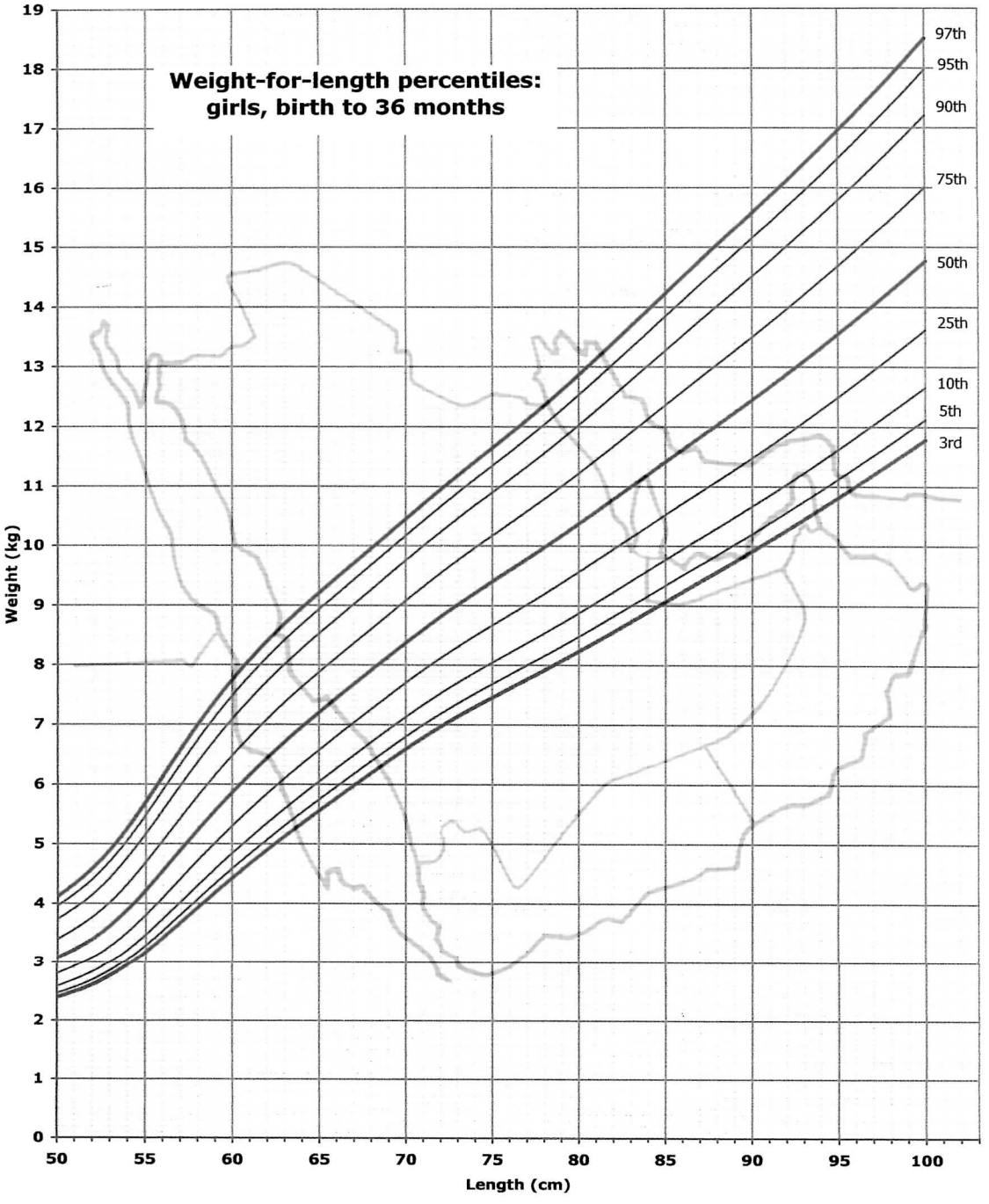

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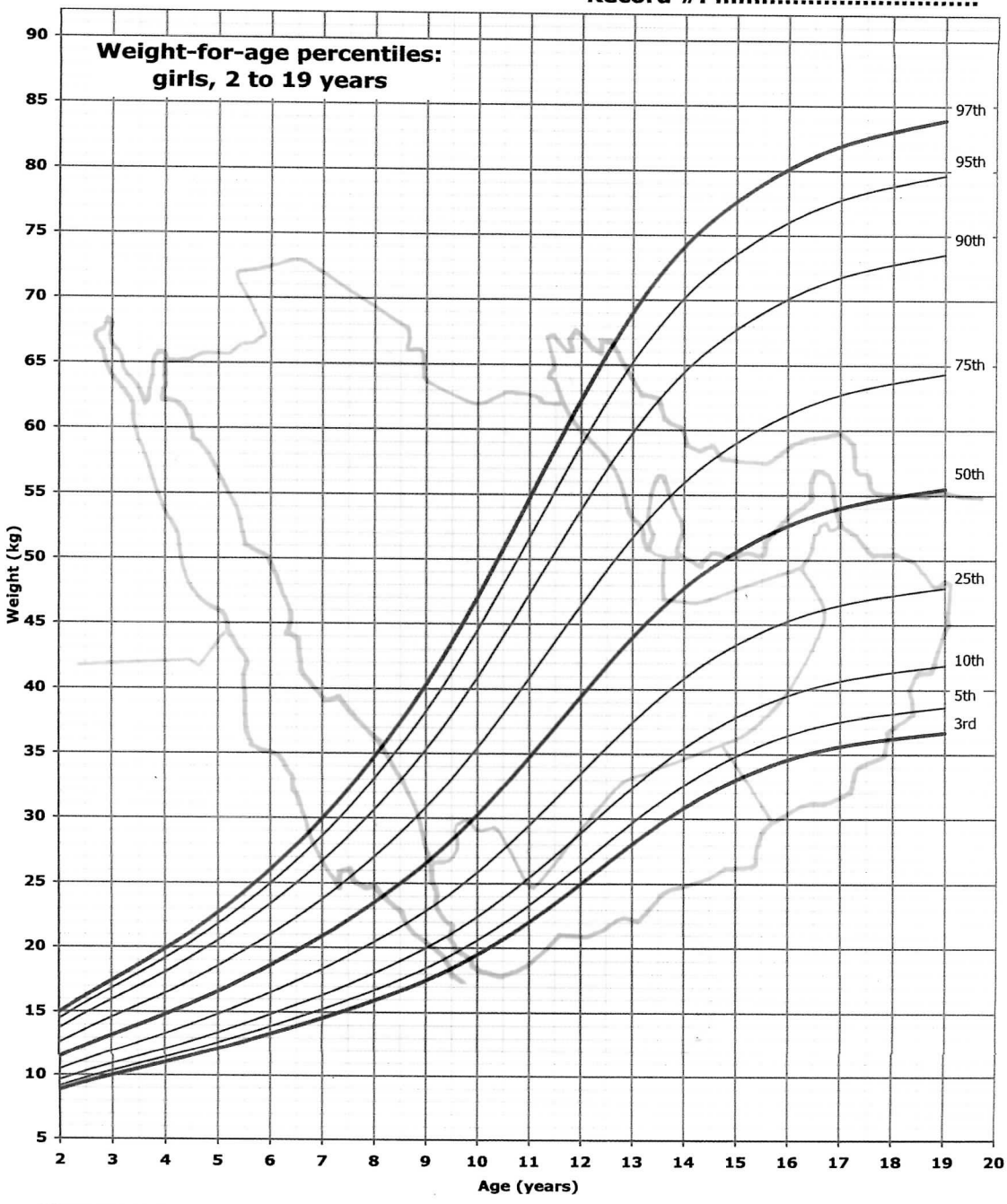

Source: Mohammad I. El Mouzan, Abdullah A. Al Salloum, Abdullah S. Al Herbish, Mansour M. Qurashi, Ahmad A. Al Omar. Health Profile for Saudi Children and Adolescents (No. AR-20-63). King Abdulaziz City for Science and Technology 2007, Riyadh, KSA.

NB: The age is based on Gregorian calender.

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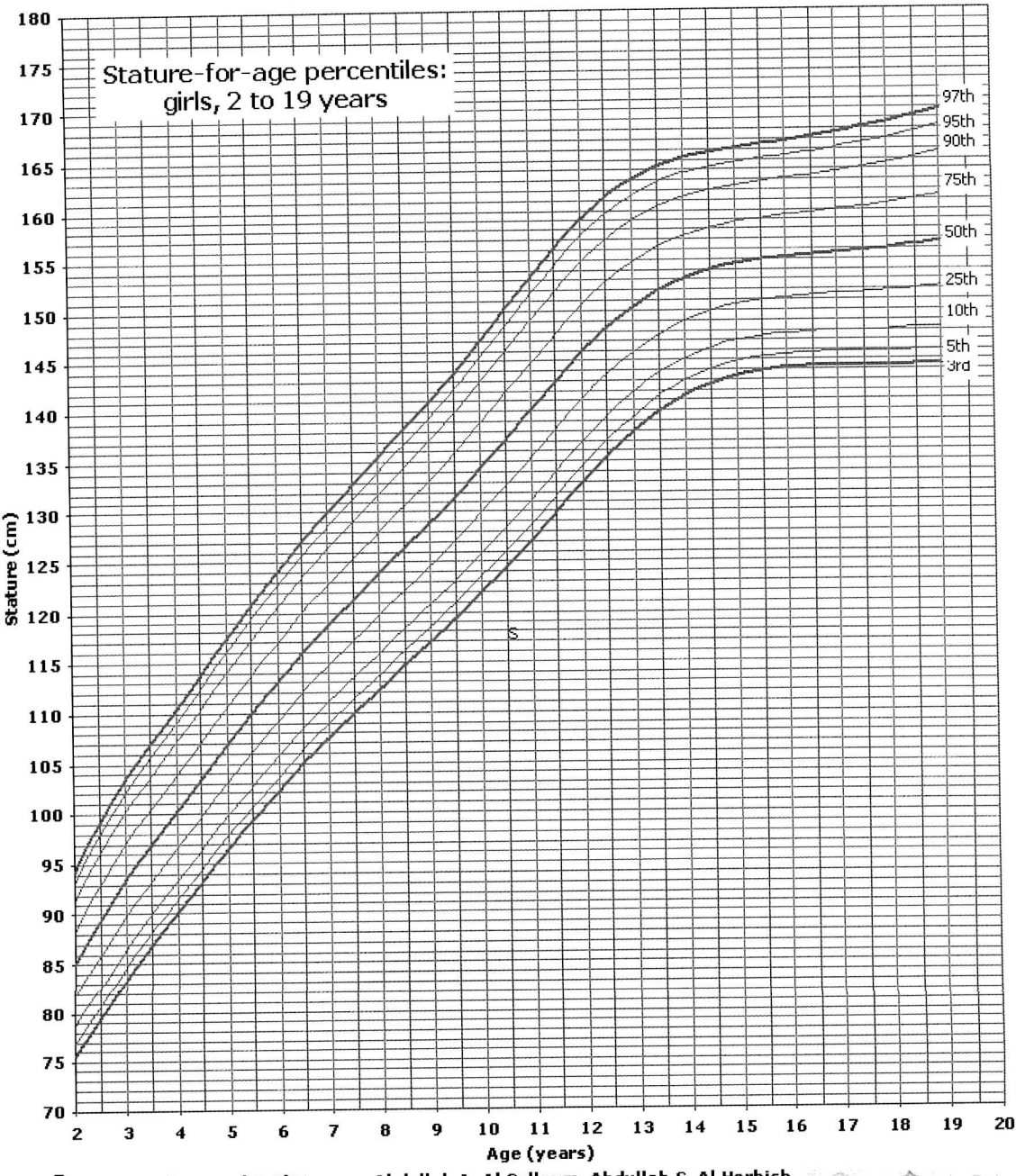

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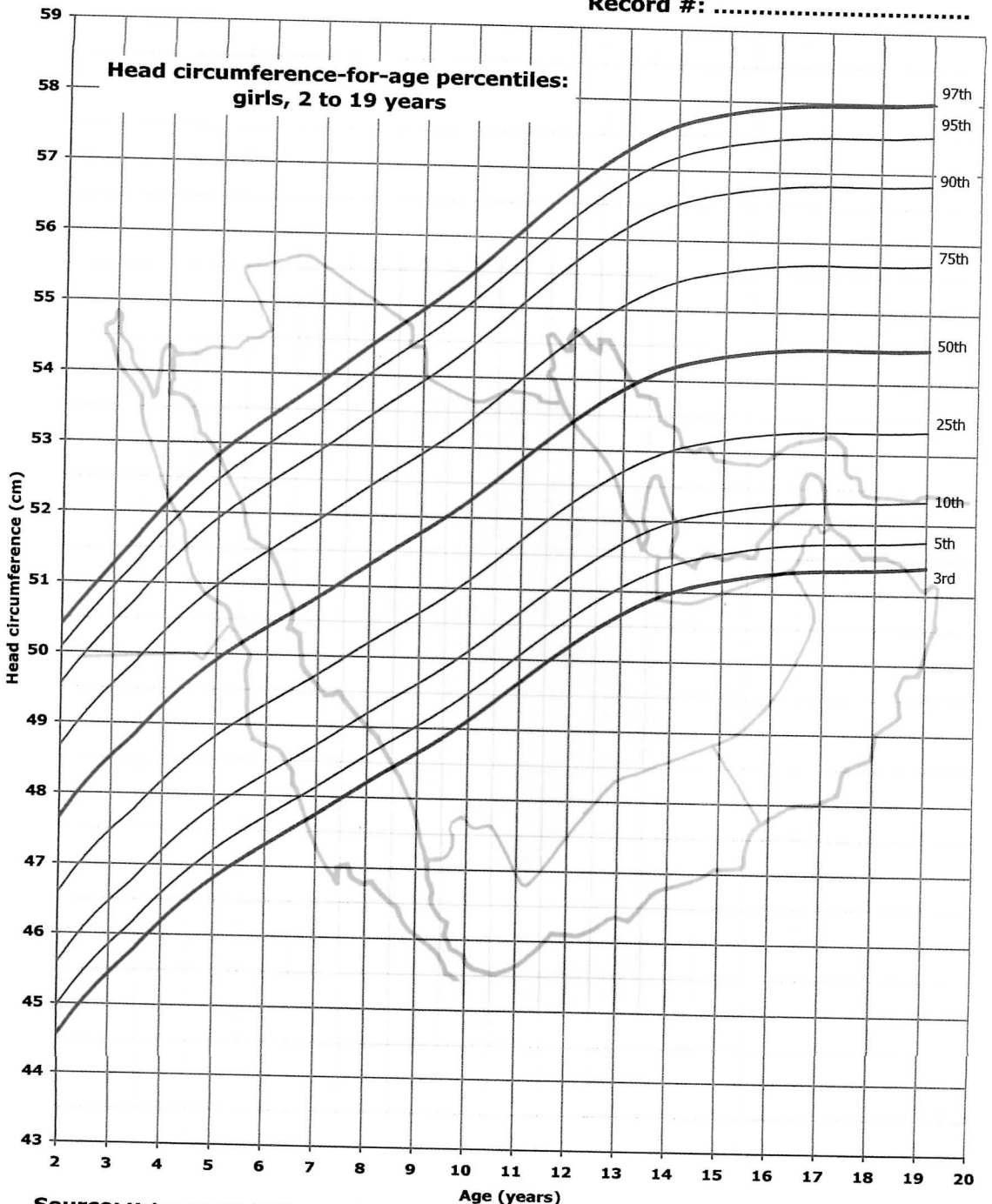

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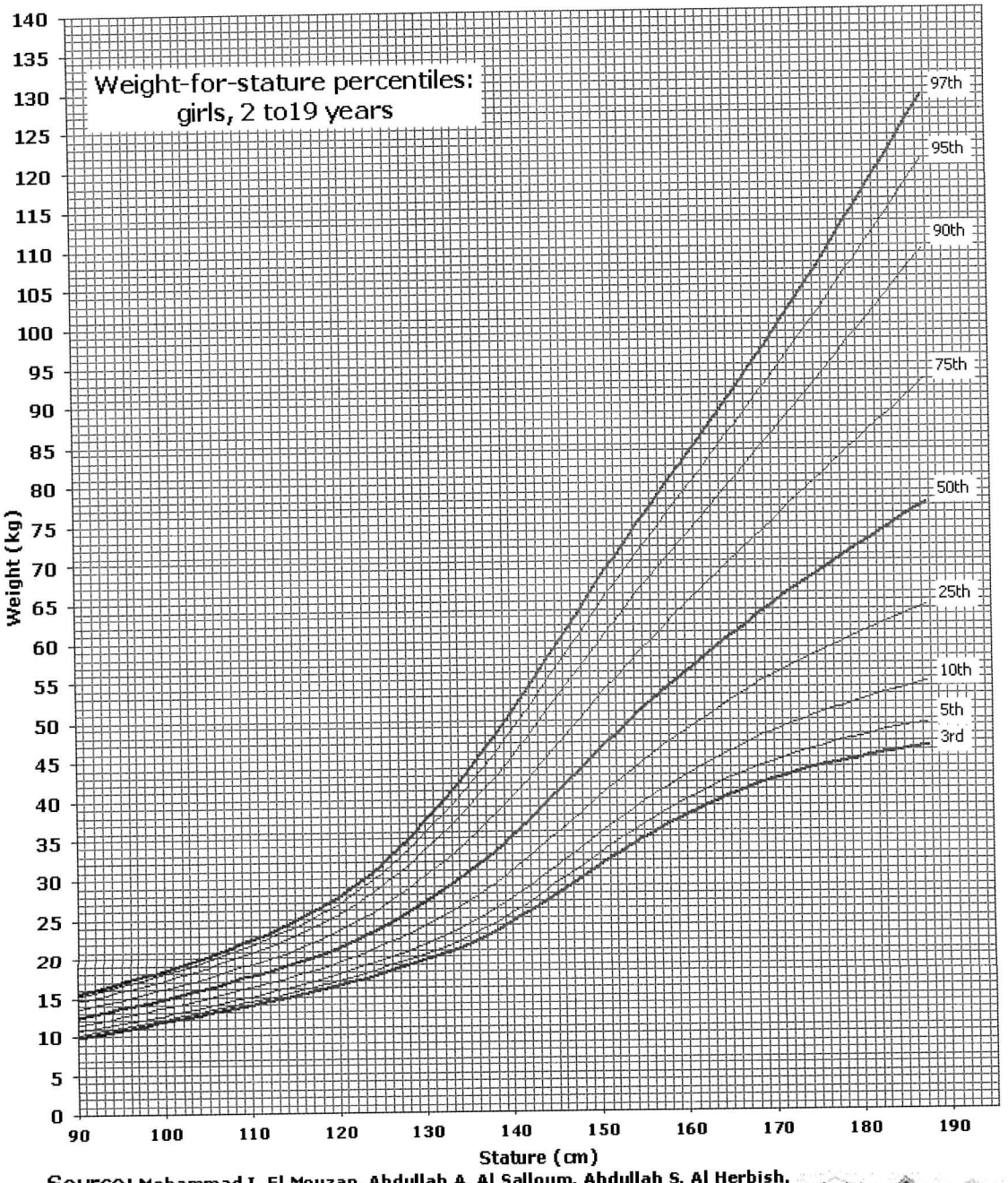

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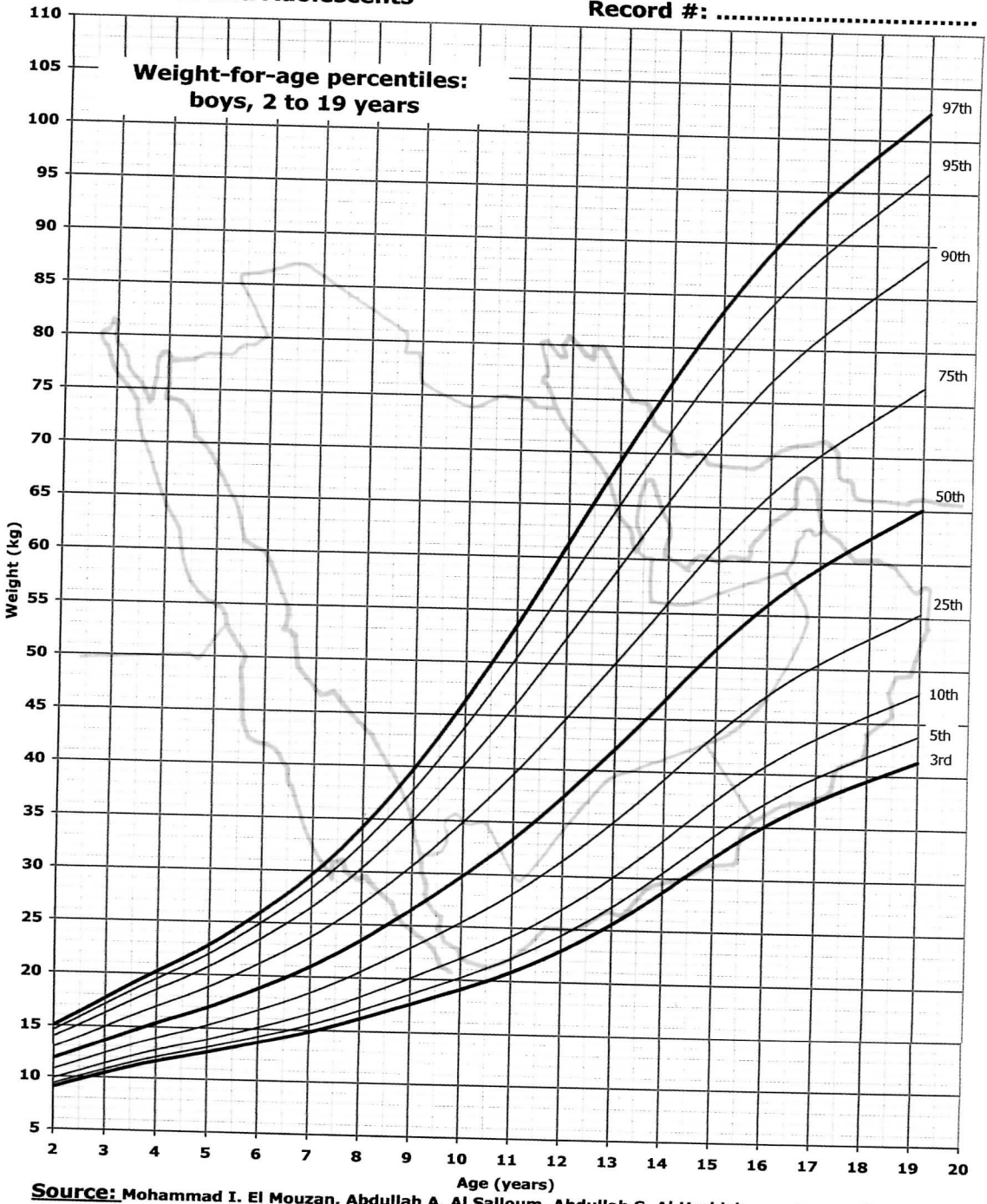

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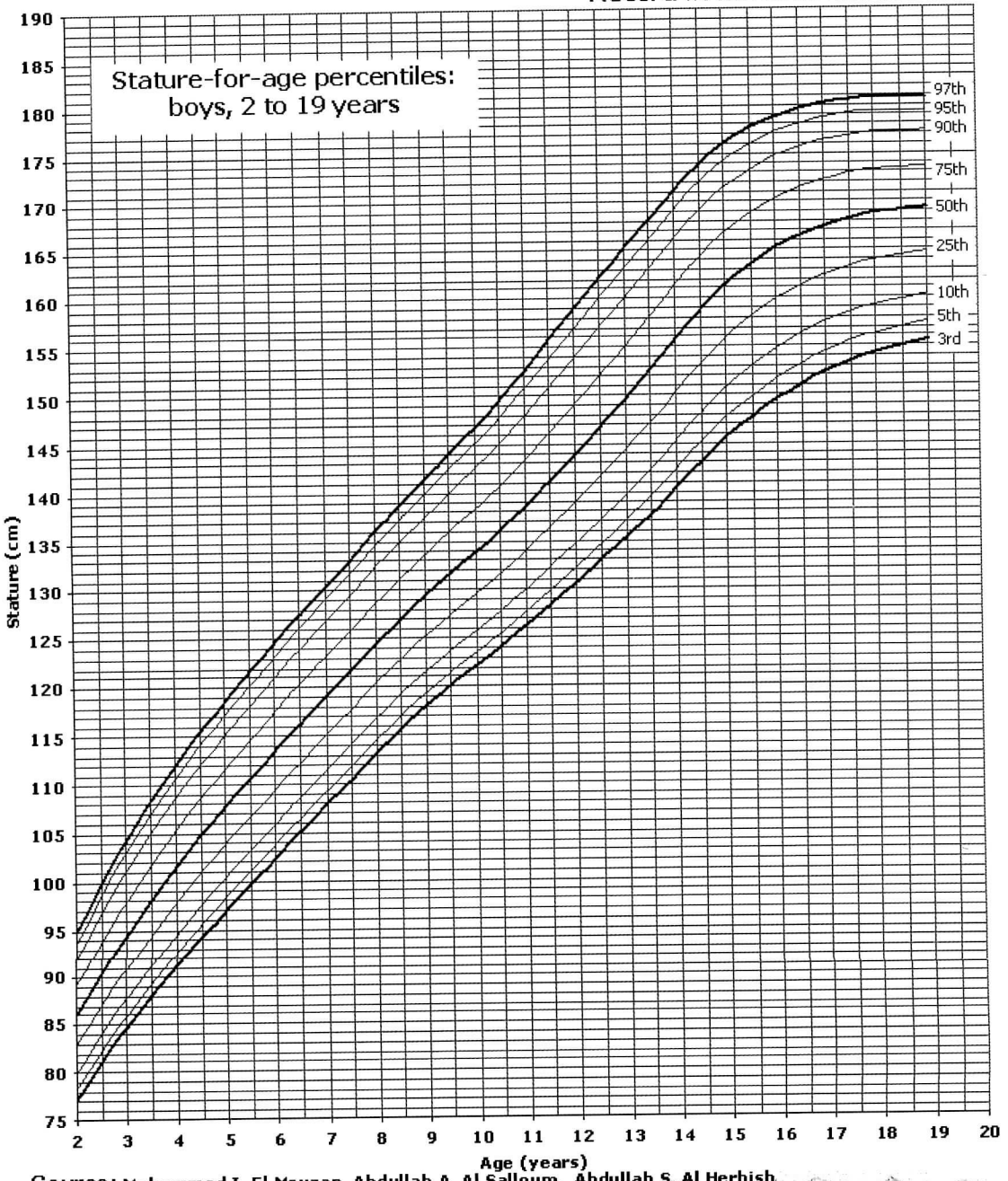

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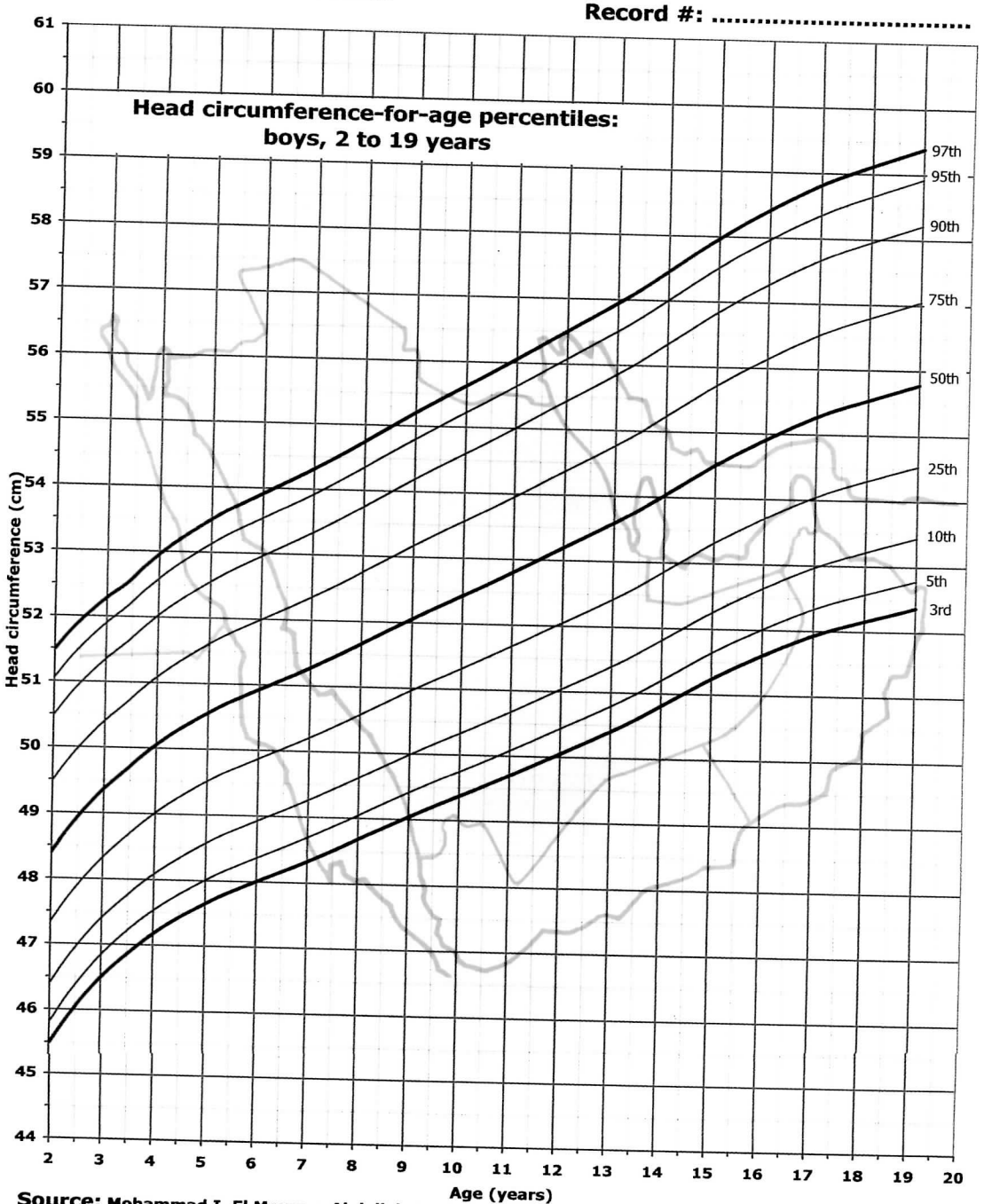

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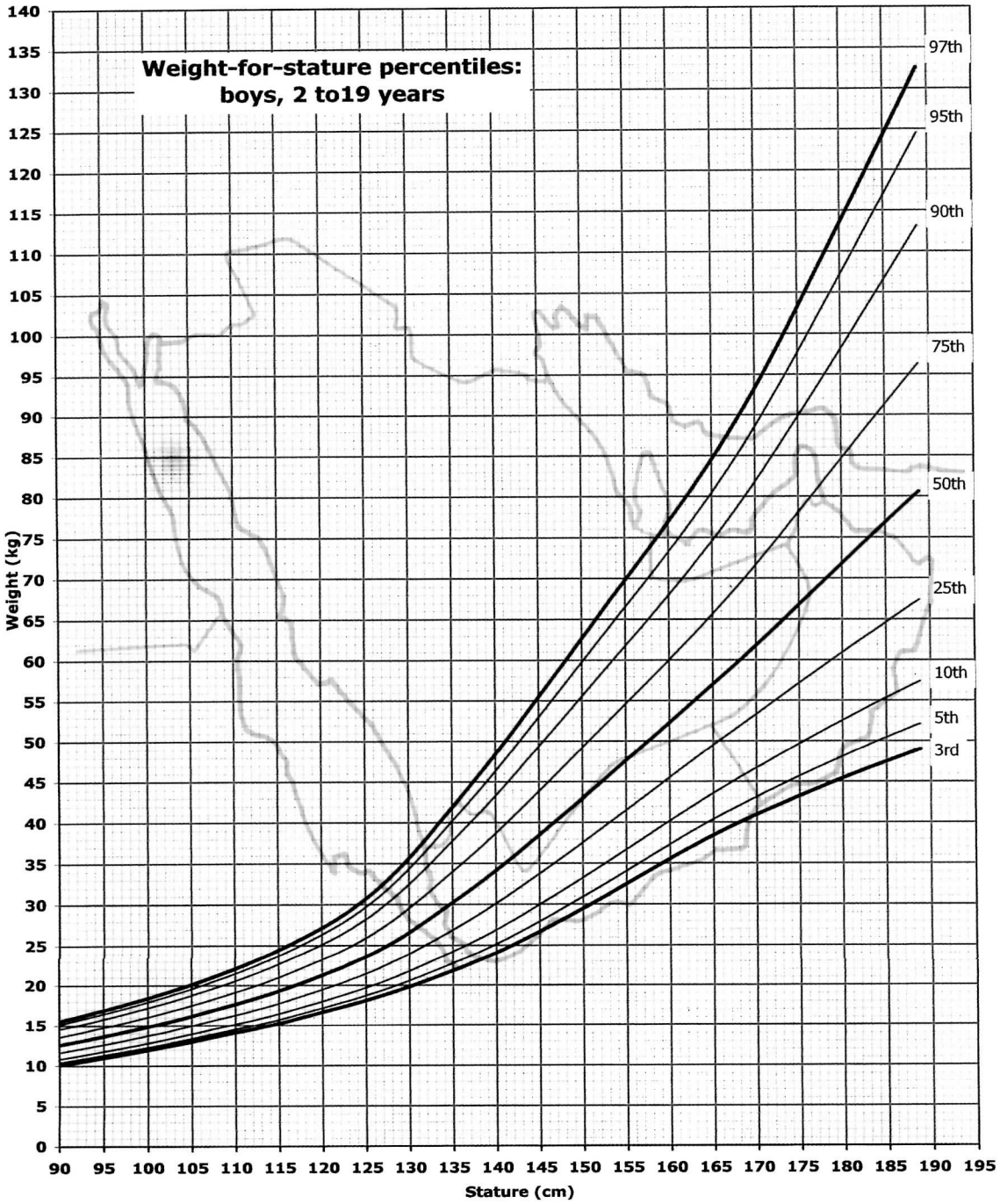

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